WIKI TEST PREP QUESTIONS

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An 8-year-old boy needs to be coaxed to go to school and, while there, often complains of severe headaches or stomach pain. His mother frequently has to take him home because of his symptoms. At night, he tries to sleep with his parents. When they insist that he sleep in his own room, he says that there are monsters in his closet. Which of the following best explains this behavior?

a. Childhood schizophrenia
b. Normal concerns of latency-age children
c. Separation anxiety disorder
d. Socialized conduct disorder
e. Symbiotic psychosis

**Correct answer**: C

Separation anxiety (option C) is a disorder of childhood that is most often seen in children 7-8 years old. The disorder manifests itself as an overwhelming fear of loss (usually of a parent) and can lead to physical complaints to avoid going to school or other activities. Separation anxiety is a normal part of development which occurs between 8-18 months; however, in a child >5 years old, separation anxiety that affects normal activity is described as a disorder.

Childhood schizophrenia (option A) develops before puberty and is presents as withdrawal, failure to develop a separate identity from the mother, and may include autistic like behaviors.

Latency is a Freudian stage of development that occurs from about age 6-puberty following the resolution of the Oedipal period. During this period the superego, the moral conscience is formed.

Socialized conduct disorder (option D) is a persistent behavior that manifests as violation of the rights of others (stealing, fighting, malicious acts). In those older than 18 years, this disorder is classified as antisocial personality disorder. Socialized conduct disorder is related to ADHD and ODD (oppositional defiant disorder).

Symbiotic psychosis (option E) is classified as a pervasive developmental disorder of early childhood described by Margaret Mahler. Although separation anxiety is feature of the psychosis, it is accompanied by developmental and social retardation.
A 30-year-old woman whose mother and grandmother have died of carcinoma of the breast refuses to have mammography. She says that she knows she is at risk but states, "I hate having my breasts squashed—it's uncomfortable." Her physician would like her to have annual mammograms. Which of the following is most likely to influence her to agree to mammography?

a. Exaggerate her risk for breast cancer
b. Insist that she obtain counseling regarding unresolved grief
c. Offer analgesia prior to mammography
d. Show her photographs of the results of untreated cancer
e. Tell her that the therapeutic relationship will be terminated unless she has annual mammograms

**Correct answer:** C

Often when patients refuse to comply with a physician’s recommendations, there is a reason for the refusal. It is important to explore the patient’s reasons for refusal. This woman has explained why she does not want a mammography. She is afraid of the discomfort involved. Therefore offering analgesia (choice C) is a reasonable first option to try to allay her fears, and have her agree to the procedure.

In general, fear tactics (choice A and D), and ultimatums (choice B and E) will not be the correct answer on the USMLE Step 1 exam.
A 52-year-old woman is admitted to the hospital because of breast cancer metastatic to the liver. Her prognosis is poor. She begs her husband to stay with her at the hospital because she is afraid to be left alone. Which of the following defense mechanisms best explains her behavior?

- a. Denial
- b. Displacement
- c. Regression
- d. Repression
- e. Sublimation

**Correct answer:** C

This patient has regressed (choice C) because she is dealing with her problem in a way a child would, and she has therefore set back her maturational clock. This defense mechanism is seen in very sick patients as well as in children under stress who may begin wetting the bed.

Denial (choice A) is the avoidance of awareness of some painful reality, such as the denial of a newly diagnosed cancer.

Displacement (choice B) is a process whereby avoided ideas and feelings are transferred to some neutral person or object. An example of this is a wife yelling at her husband because she is angry at her boss.

Repression (choice D) is an involuntary withholding of conscious awareness.

Sublimation (choice E) is the only mature defense mechanism listed in the answer choices, involving replacement of an inappropriate desire with a desire or action that is considered appropriate, differing from displacement where the desire is replaced or transferred, but to a person or object that is still inappropriate. An example of sublimation would be a person who utilizes their frustration or anger as competitiveness in sports.
A 10-month-old girl cries when her mother tries to put her in her grandmother’s arms. She happily allowed herself to be held when her grandmother visited 2 months ago. Which of the following best explains this behavior?

- a. Development of problems with socialization
- b. Maltreatment by the grandmother
- c. Sensation of a poor relationship between the mother and the grandmother
- d. Separation anxiety disorder
- e. Stranger anxiety

**Correct answer:** E

Between the ages of 7-9 months, an infant develops stranger anxiety for individuals who are not primary caregivers. Stranger anxiety is a normal developmental milestone. It manifests as crying and physical discomfort when not in the presence of a primary caregiver, and usually resolves by 24 months. It is commonly confused with the pathologic entity of separation anxiety disorder, a condition usually seen beyond the age of two years and classified as three or more of the following DSM-IV criteria:

- Recurring distress when separated from the subject of attachment (such as the mother or home)
- Persistent, excessive worrying about losing the subject of attachment
- Persistent, excessive worrying that some event will lead to separation from a major attachment
- Excessive fear about being alone without subject of attachment
- Persistent reluctance or refusal to go to sleep without being near a major attachment figure, like a mother
- Recurrent nightmares about separation
A 29-year-old woman comes to the physician for a consultation 1 month after her 7-year-old daughter was killed in a motor vehicle collision. The patient is upset and restless and wrings her hands frequently. She cannot sleep at night, has lost her appetite, and cries easily and frequently. She is preoccupied with thoughts of her daughter and sometimes thinks she momentarily sees her daughter sitting in the living room. She says she wishes that she had been hit by the car, too. She denies any thoughts of killing herself. Which of the following is the best explanation for these findings?

a. Dysthymic disorder
b. Major depressive disorder
c. Normal grief reaction
d. Obsessive-compulsive disorder
e. Schizoaffective disorder

**Correct answer: C**

Normal grief reaction, an emotional response to loss lasting up to 6 months, is characterized by feelings of physical pain, distress, and physical and emotional suffering. It may include symptoms such as diminished appetite, difficulty sleeping, restlessness, anhedonia, auditory and visual hallucinations, and feelings of guilt.

Although patients suffering from a normal grief reaction may be depressed, the diagnosis of major depressive disorder is reserved for a small subset with generalized feelings of hopelessness, helplessness, worthlessness, and guilt that persists for more than 6 months.

Dysthymic disorder has some features of major depressive disorder (anhedonia, sleep disturbance, etc.) but usually does not last as long and is not as disabling.

Obsessive-compulsive disorder is a mood disorder that involves distressing, intrusive thoughts (obsessions), and the ritualized repetitive behaviors (compulsions) sometimes employed to ward them off.

Schizoaffective disorder comprises features of both schizophrenia and a mood disorder, and may include delusions, disorganized thinking and/or speech, and manic and/or depressive episodes.
An 80-year-old man is admitted to the hospital for treatment of a burn that covers 20% of his total body surface area. Two days after admission, his behavior has changed. He accuses the staff of torturing him. He cannot recall why he was admitted to the hospital and is not oriented to date and place. His wife says he was "fine" before the burn. Which of the following is the most likely diagnosis?

- a. Adjustment disorder
- b. Delirium
- c. Paranoid personality disorder
- d. Schizophrenia, paranoid type
- e. Senile onset of dementia, Alzheimer type

correct answer: B

This man is exhibiting the classic signs of (B) delirium: an acute change in mental status, inattention, disorganized thinking, and a fluctuating level of awareness. Delirium can be caused by any of the following in the mnemonic P. DIMM WIT: Postoperative state, Dehydration/malnutrition, Infection, Medications, Metals (heavy metals exposure), Withdrawal from alcohol/drugs, Inflammation, and, as in this case, Trauma/Burns.

The acute onset of this change in mental status makes (B) delirium a much more likely diagnosis than (A) adjustment disorder, (C) Paranoid personality disorder, (D) Schizophrenia, or (E) Alzheimer's dementia.
A 30-year-old woman comes to the emergency department because she thinks she has had a heart attack. One hour ago, she had the sudden onset of chest pain, faintness, pounding heart, flushed skin, and nausea that lasted 20 minutes. She now feels better. She has limited her activity because she has had two similar episodes over the past 2 weeks. Medical evaluation is normal. Which of the following is the most appropriate nonpharmacologic therapy?

- a. Assertiveness training
- b. Cognitive behavioral therapy
- c. Dynamic psychotherapy
- d. Psychoanalysis
- e. Psychodrama

**Correct answer:** B

The woman experienced a panic attack, which can mimic heart attacks. After the resolution of a panic attack, the medical evaluation would be normal, which distinguishes these attacks from multiple heart attacks. The most appropriate nonpharmacologic therapy is cognitive behavioral therapy. The goal of this therapy is to change the way a patient interprets the world in order to change the patient’s response to problems. For patients who experience panic attacks, cognitive behavioral therapy can help them to increase adaptive behavior and decrease emotional distress, thereby decreasing the amount of panic attacks.

Assertiveness training is a subtype of cognitive behavioral therapy. However, it is not the most appropriate therapy since it only address one aspect of the woman's disorder. Assertiveness training may help decrease social anxiety, but if that is not the only trigger for this woman's panic attacks, then it would not be sufficient for therapy.

Dynamic psychotherapy is a form of talking and relational therapy that is best used for adjustment, personality, depressive, and eating disorders. It also takes several months to years for the therapy to be effective. Therefore, it would not be appropriate for helping the woman with her recurrent panic attacks.

Psychoanalysis is a specific type of treatment where the analyst, upon hearing the thoughts of the patient, formulates and then explains the unconscious basis for the patient’s symptoms and character problems. It can be used for panic attacks and anxiety disorders, but it would not be the most appropriate therapy since this woman would benefit more from relaxation training and systemic desensitization that are associated with cognitive behavioral therapy.

Psychodrama is a kind of group therapy in which group members act out their problems in a way to better understand conflicts between people. This type of therapy would not be appropriate for the woman and may even provoke another panic attack.
A 10-month-old girl is brought to the physician because of a 2-day history of diarrhea. She can sit unassisted and has started to crawl. Her mother is concerned because she babbles most of the time she is awake, and she becomes very upset if her mother leaves the room, "even for just a second." Which of the following best describes the girl's development?

a. Delayed cognitive, delayed social, delayed motor
b. Delayed cognitive, delayed social, normal motor
c. Normal cognitive, delayed social, delayed motor
d. Normal cognitive, normal social, delayed motor
e. Normal cognitive, normal social, normal motor

**Correct answer:** H

A 10-month-old child that can sit unassisted and has started to crawl has normal motor function. Infants do not start to walk until about 15 months. Her social and cognitive development are also normal. It is normal for infants to babble at 10 months and to be able to speak a few words by 15 months. Finally, the separation anxiety is also normal for infants between the ages of 10-15 months old.

If this 10-month-old girl was not able to sit unassisted, crawl, and babble, then her motor and cognitive development would be described as delayed because these are important milestones that occur in infants between 7-9 months. In addition, the presence of separation anxiety is normal for her age and up to 15-months-old and therefore is not a sign of delayed social development.
A 95-year-old woman in a nursing home has had advanced vascular dementia, severe dysphagia, and a 9-kg (20-lb) weight loss over the past 2 months. Her four children are divided regarding the decision to provide artificial feeding through a gastrostomy tube. There is no living will. The oldest son approaches the physician after a family meeting and says, "You should simply decide what is best for her and tell the others that's what we should do." Assuming the physician proceeds in this manner, which of the following best describes the physician's action?

a. Paternalism  
b. Preserving fairness in use of resources  
c. Protecting patient autonomy  
d. Rationing care  
e. Truth-telling  

Correct answer: A  
Paternalism is when a physician places his/her values system as the primary values system to make decisions for the care of the patient instead of deferring to the patient's values or autonomous decisions. In this case, the wishes of the patient should be represented by the family members as there is no living will or advanced directives.

Preserving fairness would involve ethical decisions about who should receive scarce resources; a classic example deals with who should have priority in receiving organ transplants.

A good example of protecting patient autonomy is the process of informed consent before procedures or experiments.

Rationing care acknowledges scarce health resources must be appropriated based on their cost-effectiveness, even though an individual patient's health options may be diminished.

A good example of truth-telling would involve the disclosure to a patient of his/her prognosis.
There are many causes of preventable, premature deaths in adults in the US. Of the leading causes, which can be most effectively reduced using behavioral strategies?

a. Diabetes mellitus  
b. Heart disease  
c. Lung cancer  
d. Unintentional injuries  
e. Violence and homicide

**Correct answer:** C  

Up to 50% of premature deaths in the US and Canada can be attributed to long-term patterns of behavior in living and adapting. Of the answers listed above, lung cancer (Answer C) can be significantly reduced by eliminating cigarette smoking behaviors. Behavioral strategies to eliminate smoking habits would significantly reduce the number of premature deaths from lung cancer.

While behavioral modification strategies may also reduce the number of premature deaths from diabetes mellitus (Answer A), heart disease (Answer B), unintentional injuries (Answer D), and Violence and homicide (Answer E), lung cancer has been shown to be the most directly and significantly affected by behavioral change.
A 6-year-old child is brought by ambulance to the emergency room following an automobile accident. She is covered with blood and unconscious secondary to hemorrhagic shock. Her parents urge the physician to do everything possible, but implore that no blood products be used to treat their daughter. What is the best course of action for the physician to take?

a. Administer blood without delay and contact child protective services
b. Convene an urgent meeting of the hospital ethics board
c. Engage the parents in a discussion of why they hold these beliefs
d. Transfuse blood as needed, explaining the situation to the parents
e. Treat the patient without using blood products, per the request of her parents

**correct answer:** D

In this emergent situation, the patient requires urgent blood transfusion. As a general ethical and legal rule, parents cannot withhold life- or limb-saving treatment from their children, even for religious reasons. This fact should be quickly explained to the parents, and the patient treated appropriately.

Administer blood without delay and contact child protective services (choice A) is incorrect because there is no indication that the patient has been abused. Although the parents’ request to avoid blood cannot be followed, making the request is not grounds to contact authorities.

Convene an urgent meeting of the hospital ethics board (choice B) is incorrect because a legal and ethical standard has been set for this type of situation, as described above. Further, this choice is logistically unfeasible; the patient’s condition requires immediate action.

Engage the parents in a discussion of why they hold these beliefs (choice C) may be appropriate in a non-emergent situation, but not at this point.

Treat the patient without using blood products, per the request of her parents (choice E) is incorrect. As a general rule, parents cannot withhold life- or limb-saving treatment from their children, even for religious reasons. If the patient were conscious and made the same request, it still would not be honored, as she is a minor and therefore legally incompetent.
An 18-year-old girl visits her college student health office for a required physical. She complains of irritableness and concerns about decision-making now that she has left home. On exam she appears exceedingly thin, with decreased muscle mass and dry skin. She is hypotensive, with resting bradycardia.

What other finding is most likely to be present?

a. Calluses on her hands/fingers  
b. Dental erosions  
c. Hirsutism  
d. Primary amenorrhea  
e. Secondary amenorrhea

**Correct answer:** E

This patient appears to be suffering from anorexia nervosa, as evidenced by weight loss with decreased muscle mass. Decision-making concerns, irritability, and dry skin are three of many common complaints.

Anorexia nervosa will often cause a secondary amenorrhea (choice E); treatment of anorexia nervosa should restore ovulation.

Calluses on hands/fingers (choice A), as well as dental erosions (choice B) are often seen in patients with bulimia, as a result of induced vomiting.

Hirsutism (choice C) is not typically found in patients with anorexia nervosa; on the contrary, these patients often show signs of hair loss, presumably as a result of poor nutrition. In some patients with anorexia nervosa, a thin, soft hair called lanugo grows on the body to serve as insulation in place of insufficient subcutaneous fat. This is distinct from hirsutism, however, which is characterized by coarse hair in an androgenic pattern.

Primary amenorrhea (choice D) is defined as failure of menses to occur by age 16. If present, this condition should have been worked up well before the current patient visit.
A 15-year-old girl presents to a clinic requesting advice on contraception. Which of the following is the most appropriate next step for the physician to take?

- a. Apologize that you cannot help the patient since she is a minor
- b. Inform the patient’s parents; their consent is not necessary
- c. Instruct the patient to return to clinic with her parents
- d. Obtain consent from the patient’s parents
- e. Provide the patient with counseling and/or contraception without informing her parents

**Correct answer:** E

Physicians can provide counseling or contraception to pediatric patients without their parents knowledge or consent (choice E).

The physician is not required to involve the patient’s parents (choices B,C,D) and can treat the patient despite the fact that she is a minor (choice A).
A 54 year old woman with newly diagnosed hypertension tells her physician that she does not want to take her medication because she doesn't "feel right" when taking it. Which of the following is the most appropriate response from the physician?

- a. "Are you sure it's the drug that's causing the feeling?"
- b. "I respect your autonomy to make your own decision"
- c. "The medication is necessary to prevent a stroke or heart attack"
- d. "This medication is the best option for maintaining your health"
- e. "What do you mean by not 'feeling right'?"

**Correct answer:** E

The most appropriate response in this situation is to characterize what the patient means by "not feeling right" (choice E). By gaining a better understanding of why the patient does not want to take the medication, the physician will be better equipped to help the patient, whether by treating a side effect, switching to another medication or working together to figure out another option.

"Are you sure it's the drug that's causing the feeling?" (Choice B) delegitimizes the patient's complaint without opening the door for further exploration of the "feeling".

"I respect your autonomy to make your own decision" (choice B) goes a step too far - while patients of course have autonomy and the final say in their treatment, the physician should further investigate why the patient does not want to take the medication before coming to this conclusion.

"The medication is necessary to prevent a stroke or heart attack" (choice C) is an inappropriately threatening response. Regardless of the patient's reason for discontinuing the medication, this response elicits guilt and fear rather than encouraging communication.

"This medication is the best option for maintaining your health" (choice D) similarly ignores the patient's reason for not wanting to take the medication and uses a paternalistic approach instead of a partnership.
A 47-year-old alcoholic man presents to your office with the chief complaint of “feeling depressed.” In the last few months, he had been arrested for driving under the influence and had lost his job for too many absences due to his drinking. Last week, his wife kicked him out of their house and now refuses to let him see their children. He also complains that his favorite brand of liquor must be weaker than it used to be since he finds he has to drink more of it to achieve the previously experienced effect. Presence of which one of the following favors a diagnosis of substance dependence over substance abuse?

a. Family dysfunction
b. Depression
c. Legal trouble
d. Loss of employment
e. Tolerance

correct answer: E

The DSM-IV uses the term substance abuse to identify a pattern of abnormal drug use leading to impairment of social, physical, or occupational functioning. In addition to impaired functioning, substance dependence is characterized by withdrawal and/or tolerance, which implies a physiologic as well as psychological and social component. Withdrawal is the development of physical or psychological symptoms after the reduction or cessation of intake of a substance. Tolerance is the need for increased amounts of a substance to gain the same effect.
A 40-year-old female complains of numbness and decreased sensation of her left arm and leg. She does not appear to be distressed at these findings and tells the physician that three weeks ago her 74-year-old father experienced similar symptoms and was found to have had a major stroke. Workup reveals no abnormalities. What is the most likely explanation for her symptoms?

- a. Conversion disorder
- b. Factitious disorder
- c. Hypochondriasis
- d. Malingering
- e. Somatization disorder

correct answer: A

This patient is most likely experiencing conversion disorder (choice A), given the lack of pathological findings to correlate with her symptoms, her recent life stressor and her indifference to her symptoms.

Patients with factitious disorder (choice B) fake symptoms in order to gain attention from health care professionals.

Patients with hypochondriasis (choice C) generally see many doctors and are fearful of a terrible underlying cause for their symptoms, not indifferent.

Malingers (choice D) fake symptoms for the purpose of financial or other gain, such as getting out of jail or missing work.

Somatization disorder (choice E) includes several chronic symptoms, including neurological, GI, pain, and sexual for which no underlying cause has been found.
A 27 year old female presents with an acute blindness in her right eye after having a major argument with her boyfriend. In the ED she is found to deny physical trauma to the eye but she insists she cannot see anything out of her right eye. In spite of this, she is not particularly concerned about her monocular blindness but notes that it has happened before after fighting with her boyfriend. The ED physician notes that her right eye is normal looking and reacts to light normally. What is the most likely diagnosis?

- a. conversion disorder
- b. factitious disorder
- c. malingering
- d. somatization disorder
- e. dissociative fugue

**Correct answer: A**

Conversion disorder is the most likely answer. Here is why: this is a young, female with an acute neurological deficit after a stressor. The fact that she is not concerned about the deficit points to "la belle indifferente" - which is almost always seen in this disorder. Contrary to popular belief, these people are not "faking it." There is no reported secondary gain in conversion, and no gain is made evident in this stem. Thus malingering is wrong. Factitious disorder implies creating symptoms or making oneself sick consciously but for the pleasure of "playing the sick role." Fugue is an amnestic state when there is recent travel and loss of one's identity. Somatization disorder will ALWAYS present with multiple physical problems in a female patient (stomach pain, headache, menstrual complaints, etc.)
You walk into your clinic and see a young woman, a 27 year old female who presents with a "cough and a cold" of 1 weeks duration. She mentions she has been feeling ill recently and that she thinks she has the flu. After you examine her and discuss with her what she likely has, she tells you that she wants to go out on a date with you. What is the most appropriate response?

a. "That is just impossible"
b. "Get out of my office"
c. "This is entirely inappropriate"
d. "As your physician I will not be able to"
e. "Ok. What is your number?"

**correct answer**: D

Never answer a question like this and say you will actually date your patient. Remember this is not correct and there is an obvious conflict of interest present. Being blunt (A, B, C) will strain future physician-patient relationships with this patient and may lead to provocation. D is the best answer.
Among physiological changes in the elderly, alterations in sleep patterns are common. Which of the following best describes changes in the sleep cycle that accompany aging in the elderly?

a. Decreased REM sleep and decreased slow wave sleep
b. Decreased REM sleep and increased slow wave sleep
c. Increased REM sleep and decreased slow wave sleep
d. Increased REM sleep and increased slow wave sleep
e. Increased REM sleep and no change in slow wave sleep

**Correct answer:** A
Sleep patterns in the elderly exhibit a decrease in both REM and slow wave sleep, accompanied by an increase in latency to sleep after going to bed. As a result, elderly individuals sleep for shorter durations and wake during the night more frequently.
A 25 year old woman returning from a 4 year stay in Africa presents with complaints of vomiting and diarrhea. On further examination, you notice that her breath smells like “garlic”. Biochemical studies reveal that she is deficient in a key compound of the Krebs cycle. A blood smear shows normal red blood cells. She denies any recent exposure to fire. Which of the following is the most likely cause of this patient’s presentation?

a. Lead  
b. Garlic  
c. Arsenic  
d. Carbon monoxide  
e. Mercury  

**correct answer:** C

This patient has been affected by arsenic poisoning. Arsenic is a water contaminant in many developing countries. It inhibits the Krebs cycle as follows: pyruvate dehydrogenase converts pyruvate to acetyl-CoA, which combines to oxaloacetate to form citrate. 5 cofactors are required for pyruvate dehydrogenase to function: pyrophosphate, FAD, NAD, CoA, lipoic acid. Arsenic allosterically inhibits enzymes that require lipoic acid. This causes accumulation of pyruvate, which leads to the gastrointestinal symptoms mentioned in the case (classically “rice water stools”). Severe toxicity cause neurological disturbances and death.

A is incorrect. Although lead can cause gastrointestinal problems, the lack of basophilic stippling of RBC makes it somewhat unlikely. Also, lead poisoning does not affect the Krebs cycle, and does not cause “garlic” breath.

B is incorrect. Although consumption of garlic can cause garlic smell in the breath, it is not known to gastrointestinal side effects. It does not affect enzymatic pathways, such as the Krebs cycle

D is incorrect. Carbon monoxide is most likely to occur as a result of exposure to fire, which is an absent feature in this case. Although CO poisoning has more widespread effects, it does not commonly affect the gastrointestinal system. It affects the cardiovascular system, causing tachycardia, hypertension, and sometimes myocardial ischemia. It also affects the central nervous system, leading to headache, dizziness and confusion. Other organs affected are the lungs, kidneys and muscles.

E is incorrect. Mercury poisoning typically causes sensory impairment (vision, hearing, and speech), disturbed sensation and a lack of coordination. Affected individuals commonly present with peripheral neuropathy (presenting as paresthesia or itching, burning or pain), skin discoloration (pink cheeks, fingertips and toes), edema (swelling), and desquamation (dead skin peels off in layers).
The unlabeled solid curve in the graph shown represents the loading behavior of normal human hemoglobin at a pH of 7.35 as a function of oxygen concentration. Which of the following labeled curves best represents the most likely change after addition of carbon dioxide?

a. Curve A  
b. Curve B  
c. Curve C  
d. Curve D  
e. Curve E

**Correct answer:** B  
Increased carbon dioxide shifts the oxygen-dissociation curve to the right, leading to an increase in oxygen unloading. Curve B is the only option that depicts this right-shift while maintaining the shape of the curve. Other factors you should know that can shift the curve to the right include higher temperature, increased 2,3-BPG, and lower pH.
A 2-day-old neonate becomes lethargic and uninterested in breast-feeding. Physical examination reveals tachypnea with a normal heart rate and breath sounds. Initial blood chemistry values include normal glucose, sodium, potassium, chloride, and bicarbonate (HCO3−) levels; initial blood gas values reveal a pH of 7.53, partial pressure of oxygen (PO2) of 103 mmHg, and partial pressure of carbon dioxide (PCO2) of 27 mmHg. Which of the following is the most appropriate treatment?

- a. Administer acid to treat metabolic alkalosis
- b. Administer alkali to treat metabolic acidosis
- c. Administer alkali to treat respiratory acidosis
- d. Decrease the respiratory rate to treat metabolic acidosis
- e. Decrease the respiratory rate to treat respiratory alkalosis

**Correct answer:** E

Tachypnea in term infants may result from brain injuries or metabolic diseases that irritate the respiratory center. The increased respiratory rate removes ("blows off") carbon dioxide from the lung alveoli and lowers blood CO2, forcing a shift in the indicated equilibrium toward the left:

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\text{CO}_2 + \text{H}_2\text{O} \rightleftharpoons \text{H}_2\text{CO}_3 \rightleftharpoons \text{H}^+ + \text{HCO}_3^- \]

Carbonic acid (H2CO3) can be ignored because negligible amounts are present at physiologic pH, leaving the equilibrium:

\[
\text{CO}_2 + \text{H}_2\text{O} \rightleftharpoons \text{H}^+ + \text{HCO}_3^- \]

The leftward shift to replenish exhaled CO2 decreases the hydrogen ion (H+) concentration and increases the pH (-log10[H+]) to produce alkalosis (blood pH above the physiologic norm of 7.4). This respiratory alkalosis is best treated by diminishing the respiratory rate to elevate the blood [CO2], force the above equilibrium to the right, elevate the [H+], and decrease the pH. The newborn does not have acidosis, defined as a blood pH below 7.4, either from excess blood acids (metabolic acidosis) or from increased [CO2] (respiratory acidosis). The baby also does not have metabolic alkalosis, caused by loss of hydrogen ion from the kidney (e.g., with defective tubular filtration) or stomach (e.g., with severe vomiting).
A 90-year-old woman presents for her routine doctor’s checkup. She complains of lethargy, weakness, and tingling sensation in her fingers and toes. Her eyesight has also deteriorated. On physical examination, her tongue is inflamed, and her conjunctivae are pale. Labs show that her haemoglobin is 9.8 g/dL, and her MCV is 115 µm³. Which of the following nutritional deficiencies does she most likely have?

a. Folate
b. Iron
c. Vitamin B2
d. Vitamin B3
e. Vitamin B12

**Correct answer:** E

This woman suffers from vitamin B12 deficiency (choice E). Signs and symptoms of this include a macrocytic anemia, glossitis, and neurologic findings (paresthesias, and optic neuropathy, and subacute combined degeneration).

Folate deficiency (choice A) also causes a macrocytic anemia, but it does not cause neurologic symptoms. Iron deficiency (choice B) causes a microcytic anemia. Vitamin B2, or riboflavin (choice C), is a cofactor in oxidation and reduction. Deficiency causes angular stomatitis, cheilosis, and corneal vascularization. Vitamin B3, or niacin (choice D), is also used in oxidation and reduction reactions. Deficiency causes pellagra, which manifests as diarrhea, dermatitis, and dementia.
A 24-year-old, thin, African man comes to your office as a new patient. He has recently moved to America and speaks little English. When you ask about previous medical history, he says that he has a problem with his blood and has frequent pain, but cannot explain any more. His sclerae are icteric. Upon questioning, he reports that his family members also have the same problem with their blood. What type of hematologic disorder should be suspected in this patient?

a. Genetic hemoglobinopathy
b. Iron-deficiency anemia
c. Prothrombotic state
d. Porphyria
e. Thalassemia

**Correct answer: A**

Hemoglobinopathies (A) are qualitative defects in hemoglobin, typically due to genetic mutations in globin genes. This leads to amino acid substitutions, which affects the structure, function, and stability of the hemoglobin. The most common mutation is that of beta-globin, leading to the HbS of sickle cell disease. This patient’s appearance, ancestry, and family history make a hemoglobinopathy most likely.

Iron-deficiency anemia (B) can be caused by many disease processes such as blood loss or inadequate dietary intake, but would be unlikely to cause systemic disease as this patient manifests.

Prothrombotic state (C) can be genetic or acquired. This patient’s presentation and appearance are not suggestive of a prothrombotic state, and his young age also makes this relatively less likely.

(D) Porphyrias can be inherited or acquired. They are caused by excessive production, accumulation and excretion of porphyrins or their precursors. These are much rarer than hemoglobinopathies, especially in Africans.

Thalassemia (E) is a quantitative deficiency of hemoglobin caused by an unbalanced synthesis of globin chains. Typically, an entire chain is deficient, either lacking an alpha-chain or a beta-chain. If the patient does not have a hemoglobinopathy, this diagnosis may be worth exploring.
X is an oncogene. Y is a tumor suppressor gene. Which of the following molecular events is commonly implicated in the pathogenesis of cancer?

a. X deletion
b. X inactivating mutation
c. Y activating mutation
d. Y amplification
e. Y deletion

**Correct answer:** E

Any event that disrupts the function of a tumor suppressor gene or enhances the function of an oncogene is usually associated with cancer. Therefore, deletion of a tumor suppressor gene (E), such as p53, is the correct answer. The other answer choices are not commonly implicated in cancer pathogenesis. On the contrary, an inactivating mutation of a tumor suppressor gene, or an activating mutation/amplification of an oncogene are common events in the pathogenesis of cancer.
A young Jewish couple brings in their 6-month-old baby to a pediatrician because they are concerned about the baby’s deteriorating movement. On physical exam, the pediatrician finds an exaggerated startle response, visual deterioration, macular cherry-red spots, and generalized muscle weakness, but no organomegaly or hearing impairment. A blood sample is taken and sent off to assess the activity of several enzymes. Which of the following enzymes is most likely to be defective in this patient?

- a. Arylsulfatase A
- b. B-glucocerebrosidase
- c. Hexosaminidase A
- d. Iduronidate sulfatase
- e. Sphingomyelinase

**Correct answer:** C

Given the patient’s ethnicity (Ashkenazi Jews have greatest prevalence of disease), rapid onset of neurologic defects, and macular cherry-red spots, the most likely diagnosis is Tay-Sachs disease (GM2 gangliosidosis). Tay-Sachs results from a deficiency in hexosaminidase A that causes accumulation of GM2 gangliosides.

Niemann-Pick disease (type A most common) also presents with cherry-red spots, as well as severe hearing impairment/deafness and hepatosplenomegaly. Niemann-Pick disease results from a deficiency in sphingomyelinase that causes accumulation of sphingomyelin.

Cherry-red spots are also found in mucolipidosis type I, GM1 gangliosidosis, and Krabbe disease.

Metachromatic leukodystrophy typically presents early in life with progressive deterioration of motor and neurocognitive function—specifically ataxia and dementia. It occurs due to deficiency of arylsulfatase A that results in accumulation of cerebroside sulfate.

Gaucher disease (Type I most common) typically presents with organomegaly, pancytopenia, and skeletal disease. It occurs due to deficiency of B-glucocerebrosidase that results in accumulation of glucocerebroside. It is also the most common lysosomal storage disease and prevalent in those of Ashkenazi Jewish descent.

Hunter syndrome is a mucopolysaccharidosis typically presents with organomegaly, dysostosis multiplex (defects in ossification), and neurocognitive degeneration with progressive mental retardation. It is caused by deficiency of iduronidate sulfatase that results in accumulation of mucopolysaccarides such as heparan sulfate and dermatan sulfate.
Norepinephrine stimulation of G proteins results in an increased intracellular concentration of calcium. This increase is not reversed by the extracellular calcium chelator EGTA. Which of the following is the most likely source of the increased intracellular calcium concentration?

a. Endoplasmic reticulum  
b. Golgi complex  
c. Mitochondria  
d. Nucleus  
e. Plasma membrane

**Correct answer:** A

To answer this question, you must know which organelles are sources of calcium available for use WITHIN the cell.

The main sources of calcium in eukaryotic cells are the Smooth Endoplasmic Reticulum (A, SER) and the mitochondria (C).

Release of intracellular calcium stores upon G protein activation follows a well established pathway: the beta-gamma subunit of a G-protein is cleaved and proceeds to activate Phospholipase C (PLC). PLC, in turn, cleaves the membrane bound phospholipid [phosphatidylinositol 4,5-bisphosphate] (PIP2) molecule into 2 components: Diacylglycerol (DAG) and Inositoltriphosphate (IP3). IP3 then binds to IP3 receptors present on the surface of the SER, which opens calcium channels and releases Ca2++ into the cytosol.

Calcium release by the mitochondria is not the direct target of the G-protein mediated calcium release.

The other answer choices are mere distractors and are NOT sources of calcium within animal cells.
A 3-month-old male infant is brought to the physician because of recurrent viral infections and rashes over his trunk. Lymph nodes are difficult to detect on physical examination; imaging studies indicate the lack of a thymus. Urine deoxyadenosine concentration is 100 times greater than normal. A peripheral blood smear shows a marked decrease in both mature B and T lymphocytes. A deficiency of which of the following enzymes is most likely in this patient?

a. Adenine phosphoribosyltransferase  
b. Adenosine deaminase  
c. Adenosine kinase  
d. Adenylosuccinate synthetase  
e. Hypoxanthine-guanine phosphoribosyltransferase

**Correct answer:** B

Severe combined (T and B) immunodeficiency, also known as SCID, is caused a purine salvage pathway deficiency of (B) adenosine deaminase. Without this enzyme to convert adenosine to inosine, there is an excess of ATP, dATP, and S-adenosylhomocysteine. These substances are toxic to immature lymphoid cells, and also prevent DNA synthesis through feedback inhibition of ribonucleotide reductase. Therefore, lymphoid cells fail either to reach maturity or to be produced in the first place, resulting in SCID.

Deficiency of (A) adenine phosphoribosyltransferase (APRT) may lead to kidney stones formed of adenine and salts.

Deficiency of (C) adenosine kinase is associated with nonspecific developmental abnormalities.

(D) Adenylosuccinate synthetase deficiency has been implicated in the development of hyperuricemia and gout.

Deficiency of (E) Hypoxanthine-guanine phosphoribosyltransferase causes Lesch-Nyhan Syndrome, characterized by self-mutilation, retardation, aggression, hyperuricemia, and gout.
A 3-month-old female infant from South America is adopted. She has been exclusively breast-fed. The only known medical history is treatment with an antibiotic for a gastrointestinal infection prior to arrival in the USA; the specific antibiotic is not known. Three days after receiving the infant, the adoptive parents note that she no longer moves her left upper and lower extremities. Radiologic examination shows an intracranial hemorrhage without signs of head trauma. Laboratory studies show anemia and prolonged partial thromboplastin and prothrombin times. Her condition is most likely associated with a deficiency in which of the following?

a. Gamma-Aminobutyric acid (GABA) decarboxylation  
b. Glutamic acid γ (gamma) carboxylation  
c. Heme synthesis  
d. Thymidine synthesis  
e. Xanthine oxidation

**Correct answer:** B

Prolonged bleeding times are commonly seen in infants who have a deficiency of Vitamin K. The sterile intestines of newborns are lacking in intestinal flora to synthesize Vitamin K, and this newborn is at particular risk, having recently undergone an antibiotic regimen. Therefore, she is likely deficient in Vitamin K. Vitamin K is necessary for the (B) γ (gamma) carboxylation of glutamic acid residues of proteins necessary for blood clotting. Vitamin K-dependent clotting factors II, VII, IX, and X are therefore deficient, resulting in prolonged PT and aPTT times. Anticoagulant Proteins C and S are also affected by the lack of Vitamin K, but the hypocoagulability is more significant in Vitamin K deficiency.
A 9-month-old girl has had two seizures in the past month. She was born at home and received no state-mandated newborn screening. She has developmental delays. Her skin is fair and her hair is a lighter color than that of other family members. Her diapers have a musty odor. Which of the following is most likely to have an increased concentration in this infant’s urine?

- a. Homocysteine
- b. Homogentisic acid
- c. Isoleucine
- d. Isovaleric acid
- e. Phenylacetic acid

**Correct answer: E**

The 9-month-old girl likely has phenylketonuria (PKU). In this disorder, there is a deficiency of phenylalanine hydroxylase or tetrahydrobiopterin cofactor, which causes a buildup of phenylalanine. State-mandated newborn screening looks for evidence of PKU to prevent the complications and developmental delays that occur in PKU when phenylalanine builds up in the body. The excess phenylalanine spills over into the urine as phenylacetic acid. The musty odor is a clue to the diagnosis. Since this is a disorder of aromatic amino acid metabolism, there is a musty odor in the diapers. Other common findings in PKU include fair skin, mental retardation, and growth retardation.

There would be an increased concentration of homocysteine in the urine in homocystinuria, which is caused by cystathionine synthase deficiency, decreased affinity of cystathionine synthase for pyridoxal phosphate, or methionine synthase deficiency. Common findings with this disease include mental retardation, osteoporosis, tall stature, kyphosis, lens subluxation, and atherosclerosis.

Increased isoleucine in the urine likely result from blocked degradation of branched amino acids due to a lack of alpha-ketoacid dehydrogenase deficiency. This disease is maple syrup urine disease and the urine ofen smells like maple syrup.

Homogentisic acid in the urine is seen in alkaptonuria. In this disease, there is a deficiency of homogentisic acid oxidase. The common finding in this disease is that urine turns dark on standing and connective tissue may be darker in color.

Increased concentration of isovaleric acid in the urine can be found in isovaleric acidemia. Patients with isovaleric acidemia may present in early neonatal life with a sudden onset and severe illness. The illness usually begins within a few days of birth with increasing lethargy and decreased feeding, which often causes dehydration and weight loss. The classic odor in this disease is an "odor of sweaty feet" that represents the accumulation in the body of isovaleric acid and related compounds.
A 70-year-old man has ecchymoses, perifollicular petechiae, and swelling of the gingiva. He has lived alone since the death of his wife 2 years ago. His diet consists primarily of cola and hot dogs. Which of the following is the most likely diagnosis?

a. Beriberi  
b. Kwashiorkor  
c. Pellagra  
d. Rickets  
e. Scurvy

**Correct answer:** E

Vitamin C deficiency (scurvy) classically presents with ecchymoses, petechiae, and gingival swelling. Vitamin C is necessary for the production of collagen; its deficiency results in a tendency towards weakened tissues and bleeding.

Beriberi is due to a deficiency of thiamine (B1). It may present as either "wet" (high-output heart failure and peripheral edema) or "dry" (mostly peripheral neuritis and paralysis). Either can also present with Wernicke's encephalopathy (ataxia, confusion, ophthalmoplegia/nystagmus).

Kwashiorkor is found in children, generally under age 4, who suffer from severe protein malnutrition. Symptoms include edema, red hair, muscle wasting, and eventual death.

Pellagra is due to niacin (B3) deficiency and classically presents with the D's - dermatitis, dementia, and diarrhea.

Rickets is due to a vitamin D deficiency. It classically presents with symptoms/signs of osteomalacia or "softening" of the long bones - bowed legs, "knocked knees," greenstick fractures, costochondral pain, and especially growth disorders.
A 4-year-old male with mental retardation, self-mutilation, and hyperuricemia is likely to have a deficiency of an enzyme involved in which of the following processes?

a. Breakdown of branched-chain amino acids
b. Conversion of homogentisic acid to methylacetoacetate
c. Degradation of galactocerebroside
d. Recycling of guanine and hypoxanthine
e. Synthesis of UMP and CTP

Correct answer: D

Hypoxanthine-guanine phosphoribosyl transferase (HGPRT) is an enzyme of the purine salvage pathway for recycling guanine and hypoxanthine (D) that is deficient in individuals with the X-linked recessive disorder Lesch-Nyhan syndrome. This disorder is characterized by excess uric acid production, which may produce symptoms of gout, mental retardation, spasticity, self-mutilation, and aggressive behavior.

Purine synthesis involves adding carbons and nitrogens to ribose 5-phosphate (R5P), which is a product of the hexose monophosphate (HMP) shunt. R5P is then converted to ribose phosphate pyrophosphate (RPPP), which is subsequently converted to 5'-phosphoribosylamine, the latter step being the committed step in purine nucleotide biosynthesis. Through a series of steps RPPP is converted to inosine 5'-monophosphate (IMP). Several of these biochemical steps involve transferring methyl groups from folate. This is important because folate analogues, such as methotrexate, inhibit DNA synthesis, especially in rapidly growing tumor cells, by inhibiting purine synthesis. Finally IMP is converted into either AMP or GMP. These last biochemical steps are also connected to biochemical reactions that involve adenosine deaminase, an enzyme that is deficient in individuals with the autosomal recessive form of SCID, and hypoxanthine-guanine phosphoribosyl transferase (HGPRT), a deficiency of which will produce Lesch-Nyhan Syndrome as described above. In contrast, a deficiency of homogentisic oxidase, which is involved in the conversion of homogentisic acid to methylacetoacetate, is associated with alkaptonuria. Abnormal degradation of galactocerebroside is seen in Krabbe’s disease, while abnormal breakdown of branched-chain amino acids is seen in maple syrup urine disease.
An angry group of medical students go on a hunger strike to protest the increase in their tuition. After 4 to 5 hours, which of the following occurs in their bodies?

- a. Decreased Ca++ in muscle, decreased glycogenolysis
- b. Decreased cyclic AMP, increased liver glycogen synthesis
- c. Decreased epinephrine levels, increased liver glycogenolysis
- d. Increased Ca++ in muscle, decreased glycogenolysis
- e. Increased cyclic AMP, increased liver glycogenolysis

**Correct answer:** E

In the presence of low blood glucose, epinephrine or norepinephrine interacts with specific receptors to stimulate adenylate cyclase production of cyclic AMP (Answer E). Cyclic AMP activates protein kinase, which catalyzes phosphorylation and activation of phosphorylase kinase. Activated phosphorylase kinase activates glycogen phosphorylase, which catalyzes the breakdown of glycogen. Phosphorylase kinase can be activated in two ways. Phosphorylation leads to complete activation of phosphorylase kinase. Alternatively, in muscle, the transient increases in levels of Ca++ (Answer D) associated with contraction lead to a partial activation of phosphorylase kinase. Ca++ binds to calmodulin, which is a subunit of phosphorylase kinase. Calmodulin regulates many enzymes in mammalian cells through Ca++ binding.
An elderly man who lives alone is brought to the emergency room in a confused state. He is found to have diarrhea and a pruritic rash. Which of the following nutritional deficiencies is most likely responsible for his symptoms?

- a. Vitamin B1
- b. Vitamin B3
- c. Vitamin B6
- d. Vitamin C
- e. Vitamin D

**Correct answer:** B

Lack of vitamin B3 (niacin) (Choice B), can cause Pellagra, which is characterized by altered mental status, dermatitis and diarrhea, and if left untreated, death.

Vitamin B1 (thiamine) deficiency (choice A) can cause Beriberi, characterized by polyneuritis, muscle wasting and/or dilated cardiomyopathy, and Wernicke-Korsakoff syndrome, characterized by anterograde amnesia.

Vitamin B6 (pyridoxine) deficiency (choice C) causes peripheral neuropathy, convulsions and hyperirritability.

Vitamin C (ascorbic acid) deficiency (choice D) leads to scurvy, characterized by swollen gums, easy bruising, and poor wound healing.

Vitamin D deficiency (choice E) is characterized by osteomalacia in adults (and rickets in children). The patient may have presented with a fracture in this case.
A researcher studying a rare genetic disease finds that patients with the disease are missing both copies of a gene. This gene encodes a protein containing seven-helix transmembrane domains. Which type of protein is this most likely to be?

a. G protein-coupled receptor  
b. MHC Class II  
c. Sodium-potassium ATPase  
d. T cell receptor  
e. Toll-like receptor

**correct answer:** A

The G-protein coupled receptors (A) all contain seven transmembrane segments. Importantly, all metabotropic receptors span the cell membrane and contain cytoplasmic and extracellular faces.

The other molecules mentioned are all membrane-bound proteins, but they do not contain seven transmembrane helices. The basic structure of MHC Class II (B) receptors, and T-cell (D) and Toll Like Receptors (E) should be familiar. They contain regions for ligand docking, but do not span the host cell’s membrane multiple times.

The Na/K ATPase (C) is a bidirectional active ion passing channel, that contains much more structural complexity than any of the other choices.
A 2-month old boy is brought to a clinic by his parents. The infant is jaundiced and weak. A peripheral blood smear reveals the presence of schistocytes in the blood. Based on several screening tests, an enzyme deficiency is diagnosed.

Which of the following enzymes is most likely deficient in this patient?

a. Argininosuccinase  
b. Citrate synthase  
c. Glycogen synthase  
d. Pyruvate kinase  
e. Xanthine oxidase

**correct answer:** D  
This infant presents with classical symptoms of hemolytic anemia, often caused by a lack of pyruvate kinase (PK) (D). RBCs are dependent on glycolysis for energy production. In pyruvate kinase deficiency, the last step of glycolysis cannot occur. This disease has various levels of severity but usually only manifests shortly after birth and under conditions of stress.

Argininosuccinase (A) deficiency leads to hyperammonemia and arginine deficiency.

There are no diseases related to citrate synthase (B) deficiency.

Glycogen synthase (C) deficiency can play a role in some glycogen-storage diseases.

Xanthine oxidase is a major factor in ischemia/reperfusion injury. It becomes an oxidase at low ATP levels (in ischemic conditions,) and generates reactive oxygen species once oxygen levels return to normal.
The control curve shown in the graph illustrates the relationship between the initial velocity of a reaction and the substrate concentration for any enzyme obeying Michaelis-Menten kinetics. Which of the following curves best represents the result that would be obtained by halving the amount of enzyme?

a. Curve 1  
b. Curve 2  
c. Curve 3  
d. Control curve  
e. Not represented above

**Correct answer:** C

In Michaelis-Menten kinetics, halving the amount of enzyme decreases the maximum velocity (Vmax) in half. The Vmax is how quickly a given amount of enzyme can work if provided with unlimited substrate, thus decreasing the amount of enzyme decreases the Vmax.

Choices A, B, and D are thus incorrect because the Vmax is not decreased.
A researcher analyzes blood samples taken from patients at a dialysis clinic to screen for inborn errors of metabolism and finds one male patient with an increased ceramide trihexoside level. Of note, the patient also has a history of heart failure, a painful stocking-glove neuropathy as well as multiple blue-red spots on his thighs and abdomen. What is the most likely diagnosis?

- a. Hurler’s syndrome
- b. von Gierke’s disease
- c. I-cell disease
- d. Gaucher’s disease
- e. Fabry’s disease

correct answer: E

Fabry’s disease is a deficiency of a-galactosidase A which results in all of the symptoms mentioned. The blue-red spots are angiokeratomas.

Hurler’s syndrome is a mucopolysaccharidosis characterized by coarse facial features (“gargoyle facies”), corneal clouding and hepatosplenomegaly. A deficiency of a-L-iduronidase results in an accumulation of heparan and dermatan sulfate.

von Gierke’s disease (type I glycogen storage disease) manifests with profound fasting hypoglycemia, hepatomegaly and an elevated blood lactate.

I-cell disease is a deficiency of N-acetylglicosaminyl-1-phosphotransferase which results in developmental delay, growth retardation, coarse facies with gingival hyperplasia and high levels of lysosomal enzymes in the blood.

Gaucher’s disease is a deficiency of B-glucocerebrosidase resulting in accumulation of glucocerebroside. This can present later in life with hepatosplenomegaly and bone marrow failure from infiltration of Gaucher cells.
A 60-year-old male surfer presents with a 1-inch mass on the right side of his forehead. A biopsy reveals squamous cell carcinoma. Which of the following is responsible for the formation of pyrimidine dimers in DNA, thus leading to squamous cell carcinoma?

a. Aflatoxin B1  
b. Epstein-Barr virus 
c. UV-B  
d. UV-C  
e. Vinyl chloride

**Correct answer:** C

Ultraviolet light, particularly UV-B (Answer C), is known to mutate DNA by creating pyrimidine dimers in DNA. It is associated with the formation of skin cancers, including squamous cell carcinoma, basal cell carcinoma, and malignant melanoma. Ultraviolet light is divided into UV-A (320 to 400 nm), UV-B (280 to 320 nm), and UV-C (200 to 280 nm).

UV-C (Answer D), although a potent mutagen, is not significant because it is filtered out by the ozone layer around the earth.

Some DNA viruses and RNA viruses are associated with the development of dysplasia and malignancy. For example, infection with human papillomavirus (HPV), especially types 16 and 18, is associated with cervical dysplasia; Epstein-Barr virus (EBV; Answer B) is associated with Burkitt’s lymphoma and nasopharyngeal carcinoma; hepatitis B virus (HBV) and hepatitis C virus (HCV) are associated with primary hepatocellular carcinoma; and HHV-8 is associated with Kaposi’s sarcoma. HTLV-I is an RNA retrovirus that is associated with the formation of a peculiar type of hematologic malignancy called adult T cell leukemia/lymphoma. These patients have malignant cells in their lymph nodes and blood. This malignancy is endemic in southern Japan and the Caribbean region.

Aflatoxin (choice A) is secreted by Aspergillus flavus and is a suspected liver carcinogen.

Vinyl chloride (choice E) is an industrial chemical used to make polymers. It has been associated with hepatic hemangiosarcomas.
A research study of 100 patients shows that their calcium levels range from 8.8-15.1 milligrams/deciliter (mg/dL), with a mean of 12.1 mg/dL. The calcium levels fall in a normal distribution, with a standard deviation of 1.0 mg/dL. Based on this study, we know that the percentage of calcium values below 10.1 is approximately

- 1%
- 2%
- 5%
- 8%
- 16%

**Correct answer:** B

In any normal distribution, standard deviations fall in the following manner:

68.2% fall within one standard deviation of the mean, and 95.4% fall within two standard deviations of the mean. With a mean of 12.1 and a standard deviation of 1.0, this means that approximately 4.6% of the calcium values lie outside of [10.1, 14.1]. Since one half of these will fall below this range, 4.6/2 = 2.3. Therefore, approximately 2% of the calcium values are below 10.1. (B)
A retrospective study is conducted on Gulf War veterans to determine the risk factors contributing to Gulf War syndrome. What statistical tool would be best to determine the most significant contributor?

- a. Anova-analysis
- b. Normal distribution
- c. Odds ratio
- d. Paired sample t-test
- e. Relative risk

**Correct answer:** C

An odds ratio analysis would be the best tool because it is preferred for case-control and retrospective studies. An odds ratio is defined as the odds of an event (in this case, a particular risk factor) occurring in an 'affected' group (those with Gulf War syndrome) over the odds of that event occurring in an unaffected group (those who did not develop Gulf War syndrome). Relative risk is generally employed in cohort studies and randomized controlled trials. Anova analyses and paired sample t-test require paired samples (not the case here).
The graph above illustrates a negative skew in a normal distribution. What is the relationship between the mean, median, and mode of this distribution?

a. mean < mode < median
b. mean < median < mode
c. median < mode < mean
d. median < mean < mode
e. mode < median < mean

**Correct answer:** B

The mode is easiest to determine— it’s the highest point on the distribution. The mean will always lie to the center of the distribution (so less than the mode in a negative skew). It turns out that the median will always be between the mean and mode (the explanation of which is beyond the scope of this explanation). Therefore, we have mean < median < mode for a negative skew and mode < median < mean for a positive skew.
An aspiring medical student opens up his Step 1 score report and is elated to find that his score is 230, a 92 on the 1-100 scale! The score report indicates that the mean on the exam was 215 and the standard deviation is 20. Assuming a normal distribution, what is the most likely percentile of the student’s score?

a. 54.3%
b. 72.6%
c. 84.1%
d. 97.7%
e. 99.9%

correct answer: B

This question tests your understanding of cumulative percentiles and standard deviations (sigma) in the Normal Distribution. You should know that 1(sigma) = 84.1%, 2(sigma) = 97.7%, 3(sigma) = 99.9%, and of course that 0(sigma) = 50%. Here, the mean is 215 and the deviation = 15 (230-215). A deviation of 15 corresponds to a sigma of .75 (deviation / std. deviation = 15/20). Therefore, the answer must be greater than 50% (0 sigma) and less than 84.1% (1 sigma), leaving only answer choices A or B. B is closer to 1 sigma than A is, so the most likely answer is B.
An inexpensive screening test for a disease is available through analysis of venous blood. The distributions of blood concentrations for persons with and without the disease are depicted in the graph. The disease is irreversible and fatal if not discovered and treated early. Which of the following letters represents the most appropriate cutoff point between normal and abnormal?

- a. Point A
- b. Point B
- c. Point C
- d. Point D
- e. Point E

**Correct answer: B**

This question involves the relation of predictive value to disease diagnosis. Choosing Point B as the cutoff will cause the test to have a relatively high false-positive rate: some people with concentrations within the normal range will test positive for the disease. However, because this disease is irreversible and fatal unless treated early, it is appropriate in this case to set a cutoff point that will not generate any false-negative results. Thus, setting the cutoff point at B will ensure that the negative predictive value of the test is 100%, at the expense of having a somewhat lower positive predictive value.

Choice A is incorrect because the false-positive rate would be so high that the test would not be clinically useful (the positive predictive value will be too low).

Choices C-E are incorrect because all these choices would give some false-negative results (the negative predictive value will not be 100%).
The incidence of chronic pulmonary disease in a community is 1.5 times greater than the national rate. If successful, which of the following interventions would provide the greatest decrease in chronic pulmonary disease in this community?

- Decrease the prevalence of cigarette smoking
- Decrease radon levels in homes
- Increase the number of people performing aerobic exercise
- Increase the rate of flu immunization
- Increase the rate of pneumococcal immunization

**Correct answer:** A

COPD describes patients with chronic bronchitis and/or emphysema who have a decreased forced expiratory volume. The overwhelming majority of patients with chronic bronchitis are smokers, up to 90% of all cases. Cigarette smoking also has been shown to be the major cause of emphysema. Therefore, decreasing the prevalence of cigarette smoking would lead to the greatest decrease in COPD in the community.

Persons with COPD are advised to get the flu and pneumococcal immunizations to help prevent serious infections in patients with compromised lung function. Patients with COPD have increased frequency and severity of lung infections and this likely plays a role in the etiology and progression of COPD, but not nearly to the extent of cigarette smoking. Therefore, increasing the rate of flu and pneumococcal immunizations would not lead to the greatest decrease of COPD.

Similarly, decreasing radon levels may decrease the prevalence of COPD, but only to a smaller extent when compared with cigarette smoking.

Increasing exercise will not decrease COPD in this community, especially if they continue to smoke cigarettes.
An investigator wants to study the effects of smoking on subsequent development of lung cancer. She gathers a group of 100 smokers and 100 non-smokers and follows them for 20 years to compare rates of development of lung cancer. This investigation is an example of which type of study?

- a. Case-control
- b. Cohort
- c. Cross-sectional
- d. Meta-analysis
- e. Randomized double-blind control trial

**Correct answer:** B

This study examines whether smokers are more likely to develop lung cancer than non-smokers. It is an example of a prospective, observational, cohort study (choice B), in which a group with a risk factor (smoking) is compared to a group without the risk factor to assess whether the risk factor increases the likelihood of developing the disease.

Case-Control studies (Choice A) are observational, retrospective studies in which a group of people with a given disease (lung cancer) are compared to a group without to assess the impact of a particular risk factor.

Cross-sectional studies (choice C) collect data from a group of people at a specific point in time to assess prevalence of a particular disease and risk factors. It can be used to demonstrate associations between risk factors and diseases but not causality.

A meta-analysis (choice D) is a study that pools together data from several separate studies to achieve greater statistical power.

A randomized double-blind control trial (choice E) is used to measure the effectiveness of an intervention compared to another intervention or placebo.
A researcher is conducting a study of medical students who are in the process of studying for the USMLE Step I examination. She wishes to compare the percentage of medical students who use board review books, the percentage who use question banks and the percentage that combine the two. Which of the following is the most appropriate hypothesis test for her desired results?

a. Analysis of variance
b. Chi-square test
c. Dependent t-test
d. Independent t-test
e. Z-test

**correct answer:** B

A chi-square test is most appropriate for comparing the outcomes of studies involving two or more proportions or percentages. The researcher is attempting to compare the outcome of three percentages.

Analysis of variance (ANOVA) is used to measure the outcome of three or more means.

T-tests are used to measure the significance of difference between two means. Independent t-tests are used for two groups at a singular time point. Dependent t-tests are used to measure the difference of the same group at two different time points.

Z-tests are used to measure if the difference between the sample population and the actual population is significant.
You are conducting a research study and wish to know whether two different drugs are equally effective.

H0 [null hypothesis]: Drugs A and B are equally effective.
Ha [alternative hypothesis]: Drug B is more effective than A.

Your study uses an alpha-level of alpha=0.05. The power of the test was 0.80.

If the alternate hypothesis is actually true, what is the probability that the study will show a significant difference in efficacy between the two drugs?

a. 0.05
b. 0.20
c. 0.80
d. 0.95
e. It is impossible to determine from the information given

**Correct answer:** C

A Type II Error occurs when the alternative hypothesis is true, but the null hypothesis is not rejected. In other words, this is the ability of the test to find a significant difference. The probability of committing a Type II Error is beta. The power of a test = (1 - beta). Therefore, the chance of committing a Type II Error is 0.20. However, the chance of correctly identifying a significant difference between the two drugs is equal to the power, which is 0.80 (C).

A Type I Error occurs when the null hypothesis is true, but is rejected in favor of the alternative hypothesis. The probability of a Type I Error is also known as the alpha level of the test.
You decide to do a statistical analysis of data you have collected. When plotting the numerical values for your study on a graph, you get a curve like the one depicted above. You also find that the mean is greater than the median and the median is greater than the mode. Which one of the following best describes your statistical distribution?

a. Bimodal distribution  
b. Gaussian distribution  
c. Negative skew  
d. Normal distribution  
e. Positive skew

**Correct answer:** E

The curve depicted is that of a positive skew (choice E). This is when the tail is on the right and the mean is the largest value followed by the median then the mode.

A negative skew (choice C) is when the tail is on the left and the mean is the lowest value compared to the mode and the median.

Gaussian and normal distributions (choices B and D) are the same thing. The are a perfect distribution in which the mean, mode, and median are all equal.

A bimodal distribution (choice A) is a distribution with two humps due to two divergent means.
A study of children with congenital heart disease seeks to determine risk factors for the condition that may stem from maternal medications taken during pregnancy. The study concludes, surprisingly, that children with congenital heart disease were more likely to have mothers who ate oranges while pregnant than children without congenital heart disease. What kind of error likely led to these implausible findings?

a. Poor interrater reliability
b. Recall bias
c. Sampling bias
d. Selection bias
e. Skewed distribution

**Correct answer:** B

Mothers of children with congenital heart disease are more likely to think carefully about every little thing they ate during pregnancy than mothers with children without CHD. This is an example of recall bias (choice B), in which knowledge of the presence of CHD in her child affects the way the mother remembers her history.

Poor interrater reliability (choice A) measures whether different examiners or test administrators get the same results.

Sampling bias (choice C) occurs when the subjects who volunteer or a chosen for a study are not representative of the population being studied because of confounding reasons they may have volunteered/been chosen.

Selection bias (choice D) is not a concern in a retrospective study like this one where the outcome for the children is already known (CHD or not), but rather to a prospective study where patients are assigned to groups (to eat oranges or not).

Skewed distribution (Choice E) refers to the distribution along a curve of the results. Since the question here is categorical (yes or no - oranges or not) rather than quantitative (how many oranges), so skewed distribution would not apply.
A population distribution plot of cholesterol levels among healthy and diseased individuals is shown. In defining a cut-off value for disease screening, which of the following characteristics best describe Line 2?

a. High accuracy  
b. High sensitivity, high negative predictive value  
c. High sensitivity, high positive predictive value  
d. High specificity, high negative predictive value  
e. High specificity, high positive predictive value

**Correct answer:** B

If Line 2 is the cut-off value, all diseased individuals have cholesterol levels above the cut-off and will be identified correctly by the screening; thus, the test will demonstrate high sensitivity. Additionally, there are no diseased individuals with cholesterol levels below the cut-off value; thus, there will be few false negative results, meaning the test will demonstrate a high negative predictive value. (Choice B)

High accuracy (Choice A) would be demonstrated by the cut-off value at Line 3; that is, the test would demonstrate the fewest number of combined false negatives and false positives.

High sensitivity, high positive predictive value (Choice C) is incorrect. If Line 2 is the cut-off value, all diseased individuals have cholesterol levels above the cut-off and will be identified correctly by the screening; thus, the test will demonstrate high sensitivity. However, at this cut-off value, half of the healthy population will receive false positive results; thus, the positive predictive value is low, not high.

High specificity, high negative predictive value (Choice D) is incorrect. The highest specificity is seen at Line 4: healthy patients have will have cholesterol levels below the cut-off value, and will be identified correctly (true negatives). However, at this cut-off value, half of the diseased population will receive false negative results; thus, the negative predictive value is low, not high.

High specificity, high positive predictive value (Choice E) is incorrect. The highest specificity is seen at Line 4: healthy patients have will have cholesterol levels below the cut-off value, and will be identified correctly (true negatives). This cut-off value also demonstrates a high positive predictive value: all patients with positive results would have disease.

Adapted from: http://commons.wikimedia.org/wiki/File:Beta-Fehler.png
Which of the following is used as a metabolic energy source by erythrocytes after an overnight fast?

a. Free fatty acids
b. Glucose
c. Beta-Hydroxybutyrate
d. Pyruvate
e. Triglycerides

**Correct answer:** B

Glucose is correct because erythrocytes do not have mitochondria. Accordingly, glucose is their only metabolic energy source, regardless of fasting status.

Beta-hydroxybutyrate can be used as a metabolic energy source by muscle or renal cortex tissue after an overnight fast, or by brain tissue in a prolonged fasting state. It is never an energy source for erythrocytes.

Free fatty acids can be metabolized by beta-oxidation in the mitochondria of cardiac and skeletal muscle. They are not an energy source for the brain or erythrocytes.

During fasting or exercise, pyruvate in the erythrocyte is converted into lactate then shuttled to the liver as part of the Cori Cycle. In other tissues, the mitochondrial enzyme pyruvate dehydrogenase can use pyruvate to generate acetyl-CoA for the citric acid cycle.

Triglycerides are the storage form of fatty acids, and are primarily found in the liver and adipose tissue.
Eukaryotic cells differ from prokaryotic cells in a number of significant ways. Which of the following is present only in eukaryotic organisms?

a. DNA
b. Gene splicing machinery
c. Plasmid
d. Plasma membrane
e. RNA

**Correct answer:** B

Prokaryotic DNA is transcribed and fully translated. It lacks introns and exons; consequently, it does not require the gene splicing machinery that eukaryotic cells have.
Cells from a single population are suspended in solutions X and Y, and cell volume measurements are made. The graph shows the volume of cells in solution X (solid curve) and solution Y (dashed curve). The results indicate that solution Y, with respect to X, is which of the following?

a. Hypertonic
b. Hypotonic
c. Isosmotic
d. Isotonic
e. N/A

**Correct answer:** B

The cells expand in volume when they move from solution X into solution Y, indicating that water enters the cells. This occurs when cells are placed into a hypotonic solution: the level of solute in the cell is higher than in the surrounding solution, and the osmotic pressure created by the solute causes water to move into the cell.

Choice A is incorrect because placing cells into a more hypertonic solution would cause them to decrease in volume.

Choices C and D are incorrect because placing the cells into a isosmotic or isotonic solution would cause no change in cell volume.
The nervous system increases the force of muscle contraction in a graded fashion primarily by recruitment of active motor units and by increasing which of the following?

- a. Amplitude of action potentials of single motoneurons
- b. Duration of action potentials in single motor nerve fibers
- c. Frequency of firing of individual motoneurons
- d. Number of active end-plates on the muscle fiber
- e. Rate of synthesis of acetylcholine at the motor nerve terminal

**Correct answer:** C

There is a one-one relation between the motor neuron and muscle cells (i.e., one motor neuron for each muscle cell). There is only one active end-plate from each motor neuron per muscle cell; therefore, option D is incorrect.

In addition, the action potential in the neuron will be transferred to create a distinct action potential in each muscle innervated by the neuron. The action potential is constant in its duration and in its amplitude; therefore options A and B are incorrect because these parameters cannot be changed.

Although the rate of synthesis of acetylcholine would increase the amount released by the axon terminal, the normal amount of acetylcholine released into the neuromuscular junction is enough to excite the muscle cell beyond the action threshold potential. Therefore, increasing the rate of synthesis (option E) would not affect the force of contraction.

Increasing the frequency of firing of individual motor neurons (option C) will increase the force of contraction because with each action potential firing in the muscle cell, more calcium enters the cell. Calcium drives the force of contraction and therefore with more calcium in the cell, the stronger the force of contraction will be.
An inherited metabolic disorder of carbohydrate metabolism is characterized by an abnormally increased concentration of hepatic glycogen with normal structure and no detectable increase in serum glucose concentration after oral administration of fructose. These two observations suggest that the disease is a result of the absence of which of the following enzymes?

- a. Fructokinase
- b. Glucokinase
- c. Glucose-6-phosphatase
- d. Phosphoglucomutase
- e. UDPG-glycogen transglucosylase

**Correct answer:** C

The deficient enzyme must be common to both pathways of converting glycogen to glucose and of converting fructose to glucose to yield the observed disease. Only Glucose-6-phosphatase (choice C) is common to both pathways. Glucose-6-phosphatase is required in the conversion of Glucose-6-phosphate to glucose, which is the common last step in the pathway of converting both glycogen and fructose to glucose. Deficiency of glucose-6-phosphatase is also known as Von Gierke’s disease or Type I glycogen storage disease.

Fructokinase (choice A) converts fructose to fructose-1-phosphate—the first step in the metabolism of fructose. Deficiency in this enzyme should not effect the conversion of glycogen to glucose. Fructokinase deficiency can lead to a benign disorder called essential fructosuria.

Glucokinase (choice B) is a liver-specific enzyme that converts glucose to glucose-6-phosphate as the first step of glucose metabolism. Deficiency in this enzyme should not effect conversion of glycogen to glucose or conversion of fructose to glucose.

Phosphoglucomutase (choice D) is an enzyme that allows inter-conversion between glucose-1-phosphate and glucose-6-phosphate. Deficiency in this enzyme will impair conversion of glycogen to glucose, but will not impair conversion of fructose to glucose.

UDPG-glycogen transglucosylase (choice E) is an enzyme that facilitates the incorporation of glucose into glycogen. Deficiency of this enzyme will impair glycogen synthesis.
A new opportunistic pathogen has been isolated from the stool of HIV-infected patients with persistent diarrhea. Microscopic examination indicates that this microorganism is spherical with a diameter of 3 to 5 micrometers. The presence of which of the following suggests that this is a eukaryotic microorganism?

a. Cell membrane  
b. DNA  
c. Introns in genes  
d. Messenger RNA  
e. Ribosomes

**Correct answer:** C

Eukaryotic genes contain introns and exons. Exons contain the genetic information that is transcribed into RNA while introns are spliced out during processing. Prokaryotic genes do not contain introns.

Both prokaryotic and eukaryotic cells contain (A) cell membranes, (B) DNA, (D) mRNA, and (E) ribosomes.
A medical student wishes to study the development of neural crest cells and decides to focus on a cell involved in myelination of the peripheral nervous system. On which type of cell is she most likely focusing her studies?

a. Astrocyte
b. Ependymal cell
c. Microglia
d. Oligodendrocyte
e. Schwann cell

**correct answer:** E

Schwann cells (choice E) are derivatives of neural crest cells that myelinate single axons in the peripheral nervous system.

Astrocytes (choice A) are large glial cells with end feet that are not involved in myelination.

Ependymal cells (choice B) line the central canal of the spinal canal and ventricles.

Microglia (choice C) arise from monocytes and are migratory macrophages.

Oligodendrocytes (choice D) are small glia that can myelinate many axons in the central nervous system.
An 18-year-old college student has just eaten a large container of fried rice. What is the renal cellular mechanism that prevents this sodium load from drastically increasing his plasma osmolality?

- a. Increase in aquaporin gene expression by collecting duct cells
- b. Movement of aquaporin channels to the apical surface of collecting duct cells
- c. Receptor binding of atrial natriuretic peptide (ANP) in distal tubule cells
- d. Receptor binding of spironolactone in distal tubule cells
- e. Release of urodilatin by collecting duct cells

**Correct answer:** B

An increase in sodium intake will cause an increase in plasma osmolality, triggering the release of antidiuretic hormone (ADH), a.k.a. vasopressin. The immediate effect of ADH is movement of aquaporin channels to the apical surface of collecting duct cells (Choice B).

Increase in aquaporin gene expression by collecting duct cells (Choice A) is incorrect. Although this action is the long-term effect of ADH, an upregulation of gene expression occurs over a scale of days, not minutes.

Receptor binding of atrial natriuretic peptide (ANP) in distal tubule cells (Choice C) is incorrect. ANP is released by the atria of the heart in response to an increase in blood volume, and acts on the collecting duct cells to decrease NaCl and water reabsorption. This would not decrease plasma osmolality.

Receptor binding of spironolactone in distal tubule cells (Choice D) is incorrect. Spironolactone is an exogenous medication that effects a diuresis by antagonizing the aldosterone receptor in cortical collecting tubules. This would not decrease plasma osmolality.

Release of urodilatin by collecting duct cells (Choice E) is incorrect. Urodilatin is a peptide produced by the distal tubule collecting duct in response to an increase in blood pressure/volume. It causes local inhibition of NaCl and water reabsorption; this would not decrease plasma osmolality.
An 11-year-old girl comes into your office and tells you she has started her period. She asks you to explain the menstrual cycle to her.
You tell her that the oocyte goes through different stages as it is preparing for ovulation and fertilization. What is the term for a mature follicle in the ovary that contains the oocyte that is ready for ovulation?

- a. Corpus luteum
- b. Graafian follicle
- c. Primary follicle
- d. Secondary follicle
- e. Tertiary follicle

**Correct answer: B**

Primary, secondary, and tertiary follicles are considered immature follicles. A Graafian follicle (B) has the greatest number of LH receptors and is capable of producing hormones.
A full-term 2-week-old male newborn has cyanosis. Pregnancy and delivery were uncomplicated. His lungs are clear, and a midsystolic murmur is heard that is loudest in the left third intercostal space and associated with a thrill. Which of the following is the most likely diagnosis?

a. Atrial septal defect  
b. Bicuspid aortic valve  
c. Coarctation of the aorta  
d. Patent ductus arteriosus  
e. Tetralogy of Fallot

**Correct answer:** E

The midsystolic murmur heard best in the left third intercostal space associated with a thrill is consistent with the murmur of a ventricular septal defect (VSD). Of the answer choices above, only Tetralogy of Fallot includes a VSD. In addition to a VSD, patients with Tetralogy of Fallot also have pulmonary stenosis, right ventricular hypertrophy, and overriding aorta. These patients often present shortly after birth with cyanosis.

An atrial septal defect would present with a loud S1 and a wide, fixed split S2 on physical exam and it is unlikely that the patient would present with cyanosis.

The bicuspid aortic valve would not present with cyanosis. Many patients live with unrecognized bicuspid aortic valves until later in life with the development of aortic stenosis secondary to calcifications of the valve leaflets.

Patent ductus arteriosus likely would present with a continuous machine-like murmur.

Coarctation of the aorta is associated with elevated blood pressure in the upper extremities and decreased blood pressure in the lower extremities. If these patients present shortly after birth, they often have cyanosis of the lower extremity only.
A 3-month-old male born at 37 weeks gestation presents with difficulty feeding for 3 weeks. Physical examination reveals a loud pansystolic murmur most prominent at the left lower sternal border. ECG demonstrates left ventricular hypertrophy. Chest X-ray shows cardiomegaly. During which week of gestation did the failure resulting in this defect likely occur?

a. Week 3
b. Week 4
c. Week 7
d. Week 24
e. Week 28

**Correct answer:** C

Prior to week 7 of gestation, the left and right sides of the primitive ventricle communicate through the interventricular foramen. During week 7, the right and left bulbar ridges and endocardial cushions fuse to form the membranous portion of the interventricular septum. Failure of the interventricular foramen to close at this time results in a ventricular septal defect (VSD), the most common congenital heart disorder. A small VSD is typically asymptomatic and can be heard as a high-pitched, holosystolic murmur. Larger VSDs can cause left-to-right shunting, increased pulmonary blood flow, and pulmonary hypertension. They can present during the newborn period with failure to thrive.

During the third week of gestation (Choice A), the paired endothelial heart tubes formed by the angioblastic cords fuse in the ventral midline to form a single endocardial tube that begins to function as a primitive heart.

During the fourth week of gestation (Choice B), the septa primum and secundum divide the primordial atrium into the left and right atrium.

During the twenty-fourth week of gestation (Choice D), the primitive alveoli are formed and begin to produce surfactant.

During the twenty-eight week of gestation (Choice E), fetal hematopoiesis begins to occur in the bone marrow.
A 4-year-old boy presents to his pediatrician for evaluation of poor exercise tolerance and bluish skin discoloration. Echocardiography reveals congenital heart disease; illustration of his condition is shown.

What is the name of this abnormality?

a. Aortic valve atresia
b. Patent ductus arteriosus (PDA)
c. Patent foramen ovale (PFO)
d. Tetralogy of Fallot

correct answer: D

The illustration demonstrates the four components of the Tetralogy of Fallot (Choice D): (1) pulmonic stenosis, (2) overriding aorta, (3) ventricular septal defect (VSD), and (4) right ventricular hypertrophy. This most-common cause of cyanotic congenital heart disease varies in severity; it may present as early as birth or later in the first years of life.

Aortic valve atresia (Choice A) is incorrect; the aortic valve is formed and of normal caliber in the illustration. As it does not disrupt oxygenation of blood, this condition does not usually cause cyanosis. Of note, complete aortic valve atresia is incompatible with life.

Patent ductus arteriosus (PDA) (Choice B) is incorrect; no communication between the pulmonary artery and aorta is exhibited. Embryologically, it is a remnant of the ductus arteriosus necessary for fetal circulation. Clinically, this condition leads to pulmonary hypertension and right heart hypertrophy due to excess blood flowing through the pulmonary artery.

Patent foramen ovale (PFO) (Choice C) is incorrect; no atrial septal defect is exhibited in the illustration. Embryologically, it is a remnant of the foramen ovale necessary for fetal circulation. Though PFO increases the risk of a paradoxical venous-to-arterial embolus, it would not affect exercise tolerance and most patients are asymptomatic.

In transposition of the great vessels (Choice E), the aorta arises from the right ventricle, the pulmonary artery from the left ventricle; thus, this answer is incorrect. Unless a shunt exists to allow mixing of venous and arterial blood, this cyanotic congenital heart disease is incompatible with life.

Adapted from http://commons.wikimedia.org/wiki/File:Heart_tetralogy_fallot.svg
An infant is rushed to the ER for profuse vomiting and you are asked to evaluate him. You notice that the vomitus is bile tinged, and you become concerned about a possible congenital anomaly. What is the most likely diagnosis?

- Esophageal atresia
- Hypertrophic pyloric stenosis
- Imperforated anus
- Midgut malrotation with volvulus
- Pulmonary hypoplasia

**Correct answer:** D

There is a key distinction to be made between nonbilious and bilious vomiting in general, and in infants in particular. The presence of bile in the vomitus indicates that the point of obstruction is distal to the ampulla of Vater. Bilious vomiting in infants is midgut malrotation with volvulus until proven otherwise.

Neonates with esophageal atresia typically present with excess secretions that can lead to drooling, choking, respiratory distress, and the inability to feed.

In hypertrophic pyloric stenosis, the point of obstruction is at the pylorus and thus this typically presents with nonbilious vomiting.

The other choices do not typically present with vomiting.
A 24-year-old woman has a marked increase in the size of her uterus between 26 and 30 weeks' gestation. Ultrasonography shows a marked increase in the amount of amniotic fluid; the fetus is normal in size for gestational age. Which of the following is the most likely diagnosis?

- a. Congenital heart disease
- b. Duodenal atresia
- c. Erythroblastosis fetalis
- d. Horseshoe kidney
- e. Neural tube defect

**Correct answer:** B

Fetal swallowing reduces the amount of amniotic fluid. Congenital gastrointestinal obstructions, such as duodenal atresia (Answer B) cause a buildup of amniotic fluid, or polyhydramnios. Erythroblastosis fetalis (Answer C) occurs when maternal and fetal blood mix, leading to alloimmune hemolysis, usually against Rh antigen. In late pregnancy, most amniotic fluid is composed of fetal urine. Renal anomalies that reduce the amount of urine production, such as horseshoe kidney (Answer D), are associated with oligohydramnios. Congenital heart disease does not manifest until after birth (Answer A). Neural tube defects (Answer E) usually do not affect the amount of amniotic fluid.
Blood-based and urine-based pregnancy tests both quantitatively measure the glycoprotein hormone human chorionic gonadotropin (hCG). A diagram of a 5-day-old blastocyst within the uterus is shown. Which of the lettered structures later produces hCG, resulting in a positive pregnancy test?

a. Structure A  
b. Structure B  
c. Structure C  
d. Structure D  
e. Structure E

**Correct answer:** D

Structure D (Choice D) is the trophoblast. By embryological day 6-7, this structure differentiates into the cytotrophoblast and syncytiotrophoblast. It is the syncytiotrophoblast which produces hCG to stimulate progesterone production by the corpus luteum; this hCG will result in a positive pregnancy test by about embryological day 14.

Structure A (Choice A), an endometrial blood vessel, is incorrect. Though uterine vessels undergo conformational changes allowing them to send blood into the intervillous spaces of the placenta, they do not produce hCG.

Structure B (Choice B), endometrial epithelium, is incorrect. Though invaded by the syncytiotrophoblast during implantation, this tissue does not produce hCG.

Structure C (Choice C), inner cell mass (embryoblast) is incorrect. This structure differentiates into the bilaminar disc (epiblast + hypoblast); the embryo ultimately arises from the epiblast.

Structure E (Choice E), blastocyst cavity (blastocoele) is incorrect. This structure is merely a space, and later becomes the chorionic cavity.

A 32 year old woman gives birth to a baby boy. APGAR score was 10/10 both at 1 and 5 minutes after birth. Physical exam was normal except for a sac-like structure found on the sacrolumbar region of the baby’s back. Imaging studies reveal a fluid-filled sac with the spinal cord positioned normally. What congenital defect is most likely present in this infant?

- Spina bifida occulta
- Spina bifida with meningomyelocele
- Spina bifida with rachischisis
- Spina bifida with meningocele
- Encephalocele

**Correct answer:** D

Spina bifida is a vertebral defect that occurs when the bony vertebral arches fail to properly form. Most commonly, the defect is located in the sacrolumbar region. Spina bifida with meningocele (choice D) occurs when the meninges protrude through the defect and form a CSF-filled sac with the spinal cord remaining in normal position. Besides the visible defect, these patients may present asymptptomatically because the spinal cord is not compromised.

Spina bifida occulta (choice A) is marked by tufts of hair in the sacrolumbar region and is the least severe variation of spina bifida.

Spina bifida with meningomyelocele (choice B) occurs when both the meninges and spinal cord project through the defect to form a sac. These patients usually have neurologic dysfunction that parallels the section of cord that projects through the defect.

Spina bifida with rachischisis (choice C) occurs when the posterior neuropore fails to close, thus creating an open neural tube that lies on the surface of the back. This is the most severe form of spina bifida, and patients are usually paralyzed from the level of the defect caudally.

Encephalocele (choice E) occurs when the brain and the meninges project through a skull defect.
A mother who did not receive prenatal care gives birth to a stillborn baby with anencephaly. The distraught mother asks how this could have happened. In the development of the nervous system in utero, you recall that anencephaly is due to which of the following?

a. Dilatation of the ventricles caused by an excess of CSF
b. Failure of the anterior neuropore to close
c. Failure of the posterior neuropore to close
d. Failure of the bony vertebral arches to form properly in the sacrolumbar region
e. Failure of the bony skull to form properly in the occipital region

correct answer: B
Anencephaly occurs when the anterior neuropore fails to close during week 4 of gestation. It results in failure of the brain to develop, failure of the lamina terminalis to form, and failure of the bony cranial vault to form.

Dilatation of the ventricles caused by an excess of CSF (Choice A) leads to hydrocephalus, not anencephaly.

Failure of the posterior neuropore to close (Choice C) during week 4 of gestation results in lower neural tube defects such as spina bifida.

Failure of the bony vertebral arches to form properly in the sacrolumbar region (Choice D) results in spina bifida, not anencephaly.

Failure of the bony skull to form properly in the occipital region (Choice E) results in encephalocele, characterized by the protrusion of the brain and meninges through the resulting openings.
A 3-year-old boy presents to the emergency room with his parents who report that he has not had a bowel movement in the past 4 days. They also report increased irritability, inability to eat anything without vomiting, and reddish colored urine for the past two days. On physical exam, the boy has a large abdominal mass with abdominal pain on palpation. Abdominal imaging reveals intestinal obstruction secondary to the abdominal mass. Treatment is started with chemotherapy, radiation, and surgical resection of the tumor. The pathology report of the tumor is most likely to show what histological appearance?

- Chromaffin cell neoplasm with accumulation of epinephrine and norepinephrine
- Neoplastic cells filled with clear cytoplasm organized into round or elongated collections of cells
- Small cuboid cells with round and regular nuclei
- Spindle cells arranged into bands adjacent to parenchymal cells
- Three distinct areas including a stromal area, small tubules, and tightly packed embryonic cells

**Correct answer:** E

The patient has a Wilms Tumor that is composed of 3 distinct areas including a stromal area, an area with small tubules, and embryonic cells (choice E). This tumor usually presents between the ages of 1-3 years old and tends to be a large, bulging tumor. Additional symptoms include abdominal pain, intestinal obstruction, hypertension, and hematuria.

A chromaffin cell neoplasm with abundant epinephrine and norepinephrine (choice A) describes the histology of a pheochromocytoma. These tumors occur mainly in adults in the 4th-6th decades and are associated with paroxysmal hypertension.

Neoplastic cells filled with clear cytoplasm (choice B) describes clear cell renal carcinoma, which is unlikely given the patient's age and presentation.

Small cuboid cells with round regular nuclei (choice C) characterizes the histology of renal adenomas, which are benign tumors that rarely metastasize. Renal adenomas increase in frequency with increasing age.

The histology of mesoblastic nephroma includes spindle cells arranged into bands next to normal parenchymal cells (choice D). This type of tumor is a congenital benign neoplasm. It is usually found in the first 3 months of life and must be differentiated from a Wilms tumor. The treatment is surgical resection, but local recurrence is common.
The adult kidney develops from the metanephros which has two sources: the ureteric bud and the metanephric mesoderm. If a problem in the development of the ureteric bud occurred, which of the following structures in the adult kidney would be directly affected?

a. Collecting duct  
b. Connecting tubule  
c. Distal convoluted tubule  
d. Loop of Henle  
e. Proximal convoluted tubule

**Correct answer:** A

Of the answer choices, only the collecting duct (choice A) develops from the ureteric bud. Other structures in the adult kidney that developed from the ureteric bud include the minor calyx, major calyx, renal pelvis, and ureter.

The connecting tubule (choice B), loop of Henle (choice D), distal convoluted tubule (choice C), and proximal convoluted tubule (choice E) all developed from the metanephric mesoderm.
During fetal development, the presence of testosterone and Mullerian inhibiting factor (MIF) induces masculinization of the genitalia. Which of the following represents a homologous pair of genital structures in males and females, respectively?

- a. Bulbourethral glands; Bartholin’s glands
- b. Glans penis; Labia majora
- c. Penile urethra; Paraurethral glands
- d. Prostate gland; Glans clitoris
- e. Scrotum; Vestibular bulbs

**Correct answer:** A

The bulbourethral glands in males are homologous to the Bartholin's glands in females.

The glans penis in males is homologous to the glans clitoris in females.

The penile urethra (ventral shaft of the penis) in males is homologous to the labia minora in females.

The prostate in males is homologous to the urethral and paraurethral glands of Skene in females.

The scrotum in males is homologous to the labia majora in females.
A genetic male newborn has fully developed male sexual ducts and recognizable fallopian tubes. Which of the following processes was most likely disturbed during the embryonic period?

a. Production of estrogen by the embryonic testes
b. Production of mullerian-inhibitory substance by the embryonic testes
c. Production of testosterone by the embryonic testes
d. Response of the paramesonephric (mullerian) ducts to estrogen
e. Response of the paramesonephric (mullerian) ducts to testosterone

**Correct answer:** B

Mullerian-inhibitory substance (choice B) is a hormone secreted by the Sertoli cells of the testes during embryogenesis of the fetal male. It inhibits the development of the Mullerian ducts, which would develop into the upper vagina, uterus and cervix, and fallopian tubes, if uninhibited.

Estrogen (choice A and D) and testosterone (choice C and E) have no role in the development of the mullerian ducts.
A 26 year-old sexually active female presents to her family practitioner complaining of abdominal distension, nausea, and vomiting. Her last menstrual period was 16 weeks ago, which she attributes to her recent increase in exercise. Nevertheless, her physician requests a urine sample to measure levels of a glycoprotein secreted by which of the following structures?

a. Amnioblast  
b. Cytotrophoblast  
c. Mesonephros  
d. Syncytiotrophoblast  
e. Wharton’s jelly

**Correct answer:** D

The patient's symptoms are likely secondary to a hydatidiform mole, which characteristically features painless vaginal bleeding during the second term and extraordinarily high levels of human chorionic gonadotropin (hCG), a glycoprotein secreted by the syncytiotrophoblast.

hCG is an early marker of pregnancy, appearing 6-12 days after implantation of the blastocyst. Its levels rise exponentially during the first trimester, peak at weeks 7-10 during the luteo-placental shift, and decline gradually in the following weeks.
Your 21-year-old patient comes for her first gynecological examination. You note that she has an anomaly that is usually caused by abnormal differentiation of the Mullerian ducts during embryonic development. Normally, the Mullerian ducts develop into two systems. Which of the following is one of these systems?

a. Bulbourethral glands
d. Urethral opening
e. Uterus

**Correct answer:** E

Mullerian ducts (or paramesonephric ducts) in the female differentiate into the Fallopian tubes, the uterus, and the vagina. They are paired ducts of the embryo which run down the lateral sides of the urogenital ridge and terminate at the mullerian eminence in the primitive urogenital sinus. It is tissue of mesodermal origin.
A premature baby becomes tachypneic and tachycardic immediately after birth. Loud grunting can be heard in an effort to breathe. Posterior chest exam reveals bilaterally decreased breath sounds and dullness to percussion. Contractures of the limbs and clubbing of hands and feet are also observed. The newborn’s condition is most likely caused by which of the following abnormalities?

a. Abruptio placenta
b. Oligohydramnios
c. Placenta previa
d. Polyhydramnios
e. Tetralogy of Fallot

**Correct answer:** B

Oligohydramnios is a common cause of pulmonary hypoplasia, the reason for this newborn's respiratory distress. Pulmonary maturation cannot occur since a normal amount of amniotic fluid is necessary for the growing embryo to breathe in for distension of the lungs. The diagnosis is further cemented by clubbing of the newborn's hands and feet and contractures, both caused by the reduced amniotic fluid environment of oligohydramnios.
Genes of laterality, such as iv or inv, determine the right-left axis ("sidedness"). A mutation in a laterality gene is the most logical explanation for a child who was born with which of the following abnormalities?

- a. Bifid heart
- b. Dextrocardia
- c. Double-outlet right ventricle
- d. Persistent truncus arteriosus
- e. Transposition of the great vessels

**Correct answer:** B

Dextrocardia (choice B) is a congenital malformation of the heart in which the heart is situated on the right side of the body. These patients require ECG leads to be placed in the reverse position. The condition is usually asymptomatic.

Bifid heart (choice A) is a congenital heart defect where the heart is split, or separated into two parts.

Double-outlet right ventricle (choice C) is a congenital heart disease where both the pulmonary artery and the aorta arise from the right ventricle.

Persistent truncus arteriosus (choice D) is a cyanotic congenital heart disease where a single arterial vessel arises from the base of the heart, and splits in to the coronary, systemic, and pulmonary arteries.

Transposition of the great vessels (choice E) is also a cyanotic congenital heart disease, where the aorta arises from the right ventricle, and the pulmonary artery from the left ventricle. The pulmonary and systemic circuits are in parallel rather than in series.
A 32-year-old female complains of milky discharge from her nipples and pain with urination. Upon further questioning, she notes that she has not had a period in the past 3 months. Other than white discharge from her nipples, her physical exam is unremarkable. Her pregnancy test is negative. Her urine analysis shows microscopic blood without evidence of nitrites, leukocyte esterase, RBC casts, or WBC casts. A renal ultrasound shows a dilated left ureter. Labs are sent and are as follows:

- WBC: 11,500
- Hgb: 13.5
- Platelets: 250,000
- Ca++: 12.5
- Phosphorus: 2.0

**What other medical condition is often associated with the one most likely found in this patient?**

a. Addison's disease  
b. Diabetes mellitus type II  
c. Gastrinoma  
d. Medullary carcinoma of the thyroid  
e. Pheochromocytoma

**Correct answer:** C

This patient is mostly likely suffering from the syndrome of multiple endocrine neoplasia type I (MEN I) which typically affects the pituitary (here, prolactinoma causing galactorrhea and amenorrhea), parathyroid (here, hyperparathyroidism causing kidney stones seen by increased calcium, low phosphorus, and left hydroureter), and pancreatic neoplasms (gastrinoma, option C). MEN I have an autosomal dominant inheritance, so it is important to find out about family history.

Medullary carcinoma of the thyroid (option D) and pheochromocytoma (option E) are manifestations of MEN II and MEN III, both of which are autosomal dominant inherited. MEN II also is associated with parathyroid tumors, and MEN II is associated with mucosal neuromas.

Addison's disease (option A) is caused by adrenal atrophy, and affects all 3 layers of adrenal production (glucocorticoids, mineralocorticoids, and androgens). Patients present with hypotension, and skin hyperpigmentation (due to melanocyte stimulating hormone, a by product of ACTH production). Treatment includes both glucocorticoids and mineralocorticoids.

Diabetes mellitus type II (option B) is caused by insulin resistance. Although it is a genetic disease that tends to run in families, it is not part of the MEN I syndrome.
A 20-year-old man comes to the physician’s office for a scheduled health maintenance examination. His father died of a myocardial infarction at age 55 years. Physical examination shows a tendon xanthoma on the elbow. His serum total cholesterol concentration is 360 mg/dL. A mutation is most likely to be found in which of the following genes?

- a. apoA2
- b. apoC2
- c. apoE4
- d. LDL receptor
- e. VLDL receptor

**Correct answer:** D

This patient most likely has familial hypercholesterolemia, an autosomal dominant disorder associated with a deficiency in LDL receptors (choice D). The rise in his cholesterol concentration is most likely due to a decrease in LDL receptor-mediated endocytosis of cholesterol-rich LDL. Tendon xanthomas are a typical feature of LDL receptor deficiency. In contrast, yellow papular skin lesions, or eruptive xanthomas, are a typical feature of a mutation in the VLDL receptor gene (choice E) and apoC2 (choice B).

A mutation in apoA2 (choice A) would interfere with HDL cholesterol and would not lead to skin manifestations.

A mutation in apoE4 (choice C) would cause an increase in both triglycerides and cholesterol and there may be yellow deposits in the skin creases on the palms on physical exam.
A 28-year-old woman wishing to have a baby for the first time comes to you for genetic counseling. Her husband has no medical problems but her father has hemophilia A. She is worried about the chances of her passing this on to her children. What will you tell her?

a. Each of her sons will have a 25% chance of having hemophilia.
b. Each of her sons will have a 50% chance of having hemophilia.
c. Each of her sons will have a 100% chance of having hemophilia.
d. Each of her daughters will have a 25% chance of having hemophilia.
e. Her children have no chance of having hemophilia.

correct answer: B

Hemophilia A is the classic example of an X-linked recessive disease. Assuming her mate does not have the disease, he will pass on an X to his daughters and a Y to his sons. She will pass either her normal X or her carrier X chromosome to her child, regardless of sex. The daughters therefore have no chance of having the disease, though they may be carriers. Each son has a 50% of developing the disease, depending on which X chromosome he gets from his mom.
The first twenty amino acids in the N-tail sequence of core histone H3 is MARTKTARK STGGKAPRKQ. Epigenetics play an important role in dictating the expression of genes. Which of the following answer choices correctly pairs an amino acid with its potential epigenetic post-translation modification?

a. Arginine, methylation
b. Lysine, phosphorylation
c. Methionine, phosphorylation
d. Serine, acetylation
e. Threonine, methylation

**Correct answer:** A

Acetylation by Histone Acetyltransferase (HAT) only occurs on lysine residues. Methylation by Histone Methyltransferase (HMTs) can occur on both lysine and arginine residues, and phosphorylation by serine/threonine- and tyrosine-specific kinases occurs on serine, threonine, and tyrosine residues.
Burkitt's lymphoma is characterized by the rapid proliferation of non-cleaved B cell lymphocytes. Which chromosomal translocation is commonly found in patients with Burkitt's lymphoma?

a. t(8;14)
b. t(14;18)
c. t(14;21)
d. t(21;22)
e. Trisomy 21

**Correct answer:** A

The 8 to 14 translocation is well known to cause cases of Burkitt's lymphoma by interfering with normal function of the c-myc gene (normally present on chromosome 8). Other possibilities include t(2;8) and t(8;22).

t(14;18) causes follicular B cell lymphomas. t(14;21) and t(21;22) are examples of a Robertsonian translocations (involving chromosome 21) which cause Down Syndrome. Trisomy 21 causes Down Syndrome and is not a translocation.
A 16 year old girl presents with primary amenorrhea. On further inspection, she has broad shoulders with widely spaced nipples, webbed neck, and short stature. A chromosome analysis is pending. Which of the following congenital cardiovascular defects is associated with this syndrome?

a. Coarctation of the aorta  
b. Cystic medial necrosis of ascending aorta  
c. Patent ductus arteriosus  
d. Tetralogy of Fallot  
e. Transposition of great vessels

**Correct answer:** A

Patients with Turner’s syndrome (XO karyotype) usually develop with the listed physical characteristics. In addition, they often have coarctation of the aorta, which may not be detected until several years of age by unequal blood pressure in upper versus lower extremities. Incidentally, Turner’s is also associated with bicuspid aortic valve. Transposition of the great vessels can occur in isolation or in association with maternal illnesses such as Lupus and diabetes. Cystic medial necrosis involves degeneration of elastic fibers in the ascending aorta, associated with Marfan syndrome or Ehlers-Danlos syndrome, and predisposes these patients to aneurysms. A patent ductus arteriosus can be associated with congenital rubella infection. Tetralogy of Fallot can be associated with 22q11 syndromes like DiGeorge syndrome.
A neonate is born with low-set ears, rocker-bottom feet, micrognathia, clenched hands and a prominent occiput. Cytogenetic studies would most likely reveal which of the following?

- a. Trisomy 8
- b. Trisomy 13
- c. Trisomy 16
- d. Trisomy 18
- e. Trisomy 21

**Correct answer:** D

This is Edwards syndrome, or trisomy 18. The clenched hands, rocker-bottom feet, and micrognathia are a characteristic feature.

Trisomy 13 is Patau syndrome which is usually characterized by midline defects such as cleft lip/cleft palate and holoprosencephaly. Polydactyly and microphthalmia are characteristic features.

Trisomy 16 is the most common chromosomal cause of spontaneous abortion in utero.

Trisomy 21 is Down syndrome, which is characterized by flat facial profile, prominent epicanthal folds, single palmar crease, duodenal atresia and congenital heart disease.

Trisomy 8 frequently results in miscarriage and those that survive to term usually exhibit trisomy 8 mosaicism. This syndrome results in mental retardation, relatively specific facies, absent or dysplastic patellas, joint contractures and other anomalies.
While rounding in the neonatal nursery, a routine physical exam on an infant reveals aniridia and genital malformations. For which condition should this infant be screened?

- a. Endocardial cushion defects
- b. Hypercalcemia
- c. Hypocalcemia
- d. Tay-Sachs
- e. Wilms tumor

**Correct answer: E**

This is the WAGR complex: Wilms tumor, aniridia, genital malformations and mental retardation. It is called AGR if Wilms tumor is not present, but >30% of people with AGR will develop Wilms tumor.

Endocardial cushion defects are a common feature of Down syndrome.

Hypocalcemia is a feature of the 22q11 syndromes. DiGeorge syndrome specifically causes anomalies of the 3rd and 4th branchial pouches which lead to hypoparathyroidism and hypocalcemia.

Tay-Sachs is a defect in hexosaminidase A which leads to accumulation of GM2 ganglioside. Patients usually present with progressive developmental delay and cherry-red spot on fundoscopic exam.

Hypercalcemia is a feature of Williams syndrome, which is characterized by elfin facies, "cocktail party" personality, and other anomalies. It is a deletion in chromosome 7.
The above numbered codons relate to the partial sequence of an mRNA for a protein. Termination codons are UAG, UGA, and UAA; the initiation codon is AUG. Insertion of uridine between the end of codon 135 and the beginning of 136 will result in a protein containing how many amino acids?

a. 134  
b. 135  
c. 136  
d. 137  
e. 138

correct answer: E

Inserting a single nucleotide between codons 135 and 136 shifts the reading frame, so that the codons which follow now read: UUU (new codon 136), AUG (new codon 137), UGU (new codon 138), UAA (new codon 139), with a leftover uridine at the end. However, the new sequence includes a stop codon, UAA (new codon 139), which means that the total number of amino acids will be 138 (choice E).
A normal peptide has 100 amino acids. Following mutation, it has only 20; 1–10 are the same as the normal peptide; 11–20 differ from the normal peptide. Which of the following mutations occurred?

- a. Nucleotide deletion in the codon for amino acid 11 (codon 11)
- b. Nucleotide deletion in codon 20
- c. Nucleotide deletion in the intervening sequence
- d. Nucleotide substitution in codon 11
- e. Nucleotide substitution in codon 20

Correct answer: A
Nucleotide deletion in the codon for amino acid 11 (A) would cause a frameshift mutation. Such a mutation could result in a different amino acid at positions 11 and up, with the potential for an aberrant stop codon and early termination, as observed.

Nucleotide deletion in codon 20 (B) would cause a frameshift mutation. This mutation may result in a different amino acid 20, perhaps with termination thereafter. However, this option does not explain the difference observed in amino acids 11–19.

Nucleotide deletion in the intervening sequence (a.k.a. intron) (C) would not affect peptide sequence because introns are spliced out of the primary transcript, and thus are not translated.

Nucleotide substitution in codon 11 (D) may result in a different amino acid 11, but such a substitution would not change the rest of the nucleic acid sequence.

Nucleotide substitution in codon 20 (E) may result in a different amino acid 20, but such a substitution would not change the rest of the nucleic acid sequence.
A 3-year-old boy with mental retardation has hepatosplenomegaly. Analysis of tissue obtained on biopsy of the liver shows unusually large amounts of glucocerebroside. Which of the following enzymes is most likely to be deficient?

a. Hormone-sensitive lipase  
b. Lipoprotein lipase  
c. Lysosomal hydrolase  
d. Sphingolipid synthase  
e. Tissue phospholipase  

**Correct answer:** C

The clinical picture suggests a diagnosis of Gaucher's disease. Gaucher's disease is the most common variant of a group of diseases called lysosomal storage diseases which are caused by deficiency in any one of many lysosomal enzymes. Gaucher's disease occurs due to the deficiency of beta-glucocerebrosidase, a lysosomal hydrolase (choice C) that converts glucocerebroside to cerebroside. Glucocerebroside accumulates and can then cause the clinical picture of mental retardation and hepatosplenomegaly.

Hormone-sensitive lipase (choice A) is an enzyme found in the cytosol of adipocytes that hydrolyzes triglycerides to free fatty acids and glycerols.

Lipoprotein lipase (choice B) is an enzyme found in endothelial cells lining capillaries that hydrolyzes triglycerides circulating in chylomicrons and VLDLs into free fatty acids and glycerols.

Sphingolipid synthase (choice D) is an enzyme that uses ceramide as a substrate for sphingomyelin production.

Tissue phospholipase (choice E) is an enzyme that hydrolyzes phospholipids into free fatty acids and other lipophilic products.
A 7-month-old infant is brought to the physician’s office because of poor weight gain despite large food intake. He has had two episodes of pneumonia and has frequent bulky stools. He coughs frequently. X-rays of the lungs show increased markings and hyperinflation. Trypsin is absent in a fresh stool sample, and the fat content is increased. Which of the following is the most likely cause of this infant’s disorder?

a. Autoimmune disorder  
b. Defective ion transport at epithelial surfaces  
c. Disaccharidase deficiency  
d. Inability to synthesize apolipoprotein B  
e. Villous atrophy of the jejunum

**Correct answer:** B

This infant has the clinical presentation of cystic fibrosis, which is due to defective ion transport at epithelial surfaces (choice B). This is an autosomal recessive disease due to a mutation in the chloride transporter, cystic fibrosis transmembrane conductance regulator (CFTR). Patients present with meconium ileus, deficiencies of pancreatic enzymes, pulmonary obstruction, frequent pulmonary infection, bronchiectasis, cor pulmonale, and respiratory failure. Other findings include liver cirrhosis, infertility, and elevated NaCl concentrations in sweat.

This is not due to an autoimmune disorder (choice A), disaccharidase deficiency (choice C), inability to synthesize apolipoprotein B (choice D), or villous atrophy of the jejunum (choice E).
A 6-year-old boy has a large intra-abdominal mass in the midline just above the symphysis pubis. During an operation, a cystic mass is found attached to the umbilicus and to the apex of the bladder. Which of the following is the most likely diagnosis?

- a. Hydrocele
- b. Meckel cyst
- c. Meckel diverticulum
- d. Omphalocele
- e. Urachal cyst

**Correct answer:** E

The urachus is a connection between the dome of the bladder and the umbilicus that obliterates during development. A urachal cyst (Answer E) is usually located in between the obliterated ends of this developmental duct. A hydrocele (Answer A) is a benign collection of peritoneal or scrotal fluid in the scrotum. It is usually due to patent processus vaginalis or scrotal fluid imbalance. A meckel cyst (Answer B), aka duplication cyst, often contains ectopic gastric mucosa. It is most often located in the ileum, duodenum or stomach. A Meckel's diverticulum (Answer C) is an intestinal outpouching that usually occurs in the ileum. It occurs when the vitelline duct, which connects the primary midgut and the yolk sac, fails to obliterate. During the 6th week of development, the midgut herniates through the umbilical cord. An omphalocele (Answer D) occurs during week 12 when this hernation fails to retract, leaving an amnion-covered sac protruding through the umbilicus.
A 4-year-old boy has delayed motor development and choreoathetosis. He had normal development at birth. He chews his fingers and lips, which has resulted in tissue loss. He has arthritis. Serum and urine uric acid concentrations are increased. Which of the following abnormalities is the most likely cause of these findings?

- a. Adenine phosphoribosyltransferase deficiency
- b. Hypoxanthine-guanine phosphoribosyltransferase deficiency
- c. Increased cellular turnover of nucleic acids
- d. Increased conversion of hypoxanthine to inosine monophosphate
- e. Phosphoribosylpyrophosphate synthetase deficiency

correct answer: B

Self-mutilation is a classic symptom of Lesch-Nyhan syndrome, a genetic HPRT enzyme deficiency. Purines, namely guanine and hypoxanthine, are recycled to GMP and IMP by HPRT (Answer D). A deficiency in HPRT (Answer B) forces purines to be degraded to excess uric acid. Another purine salvage pathway deficiency, APRT deficiency (Answer A), leads to toxic buildup of adenosine metabolites in B and T cells, one of the causes of SCID. Answer C describes nucleic acid recycling, which should prevent uric acid accumulation. PRPP (Answer E) is a purine synthesis enzyme; PRPP superactivity, not deficiency, would cause uric acid buildup.
A 1-week-old newborn has microcephaly, intrauterine growth retardation, widely spaced eyes, and a cat-like cry. Which of the following karyotypes is most likely?

- a. 45,X
- b. 46,XX
- c. 46,XX,del(5)(p12)
- d. 46,XX,t(14;21)
- e. 46,XY

**Correct answer:** C

All the described abnormalities point to Cri-du-Chat syndrome, uniquely characterized by a 'crying of the cat' like vocalization made by affected infants. Cri-du-chat results from deletion of part of the short arm of chromosome 5, and spares the normal diploid number. This answer choice is 46,XX,del(5)(p12).

46,XX,del(5)(p12) describes a rare translocation that results in Acute Lymphoblastic Leukemia, not developmental disorders.

46,XX,del(5)(p12) indicates a trisomy in Chromosome 13, resulting in severe mental retardation and growth abnormalities called Patau's Syndrome. The description above suggests that this is not the best answer.

47,XX,+21 describes Trisomy in chr 21, which is classically Down's Syndrome. While Down's syndrome may result in microcephaly, it would most prominently cause mental retardation.

69,XXY is a total triploidy and usually incompatible with life.
A specific congenital form of deafness results from mutation of a sequence of connexin 26 that encodes the channel within the connexon of the gap junction. This mutation is most likely to affect which of the following domains of the protein?

a. Amino terminus  
b. Carboxyl terminus  
c. Cytoplasmic loop  
d. Extracellular loop  
e. Transmembrane region

**Correct answer:** E

A connexon is an assembly of 6 proteins (called connexins) that forms a gap junction in the (E) transmembrane region between the cytoplasm of two adjacent cells. The connexon is actually the hemichannel supplied by a cell on one side of the junction; two connexons from opposing cells normally come together to form the complete intercellular gap junction channel. However, in some cells, the hemichannel itself is active as a conduit between the cytoplasm and the extracellular space.
A 24 year old 6'9" patient who is new to your city comes for a first checkup. He has a medical history significant for two idiopathic deep vein thromboses; he tells you that his brother also had a cerebral sinus thrombosis as a child and his mother had three spontaneous abortions. He has had eye problems in the past but doesn’t recall what they were. On physical exam, the patient appears to be very tall and lanky. He does not have hyperextensible joints, a notable chest wall deformity, or small genitalia. You order some lab tests. What genetic syndrome is this patient most likely to have?

a. Marfan syndrome
b. Ehlers-Danlos syndrome
c. Kleinfelter syndrome
d. Homocystinuria
e. Antiphospholipid syndrome

**correct answer:** D
This patient is most likely to have homocystinuria. It is an autosomal recessive disease caused by a deficiency of cystathionine beta synthase. Clinical manifestations include a tendency for idiopathic arterial or venous thrombosis, which is the most common cause of death. Other manifestations include ectopia lentis, Marfanoid habitus, and cognitive impairment.

Marfan syndrome is not associated with a tendency towards idiopathic thrombosis. Both Marfan and Ehlers-Danlos are likely to present with hyperextensible joints. Small genitalia are common in Kleinfelter syndrome. Antiphospholipid syndrome is associated with idiopathic thrombosis, but not with Marfanoid habitus or eye abnormalities.
A 7-year-old male has a 3-year history of intermittent vague burning/aching of his fingers and toes. On physical examination, skin lesions are apparent which are identified as angiokeratomas. Family history is significant for an older brother who suffered from renal failure. The annual diagnosis rate for this condition is 1/200,000 males in the US population. What is the gene frequency of this X-linked recessive mutation?

a. 1/200,000  
b. 2/200,000  
c. (1/200,000) x (1/200,000)  
d. (1/400,000)  
e. (1/600,000)

**correct answer: A**

This child's presentation is classic for Fabry's disease, an X-linked recessive lysosomal storage disease. This question actually tests understanding of the Hardy-Weinberg principle. A male with Fabry's will have one copy of the disease-causing recessive mutation. As a result, the incidence on any X-linked recessive disease ("annual diagnosis rate") is a direct estimate of the gene frequency in a population.

Note:
Prevalence is the total cases in a population at a given time; while incidence refers to the new cases.

prevalence=incidence x disease duration
A laboratory worker notes that when she tries to hybridize a radioactive probe (10 nucleotides long) to a Southern blot of DNA from cultured cells, a solid, dark pattern emerges. What is the most likely reason for this pattern?

a. The temperature of the hybridization mix is too low, resulting in aberrant signals
b. The salt concentration of the mix needs to be increased
c. The cell culture is contaminated with mycoplasma
d. A longer probe is needed to increase specificity
e. The blot contains incompletely digested DNA and hence the complementary sites are not adequately separated

**Correct answer:** D

The answer is D. A probe of only 10 nucleotides (even if it does not contain an obviously repetitive sequence) will have over 1000 matches in the genome. Raising the temperature of the hybridization mix may improve the signal but eliminating competition from ∼1000 sites is unlikely (choice A). Increasing the salt concentration also can increase stringency but is not likely to eliminate the overwhelming number of competitive sites (choice B). Although the cell culture might be contaminated, unless the probe is related to the contaminating sequence(s) this will not be the problem (choice C). Incompletely digested DNA on the blot can lead to aberrant hybridization, but using a short probe is likely to give a broad smear of signals (choice E).
A patient with chorea and dementia is diagnosed with Huntington’s Disease. Which of the following correctly describes the genetic abnormality present in this patient?

- a. Disruption in HOX gene expression
- b. Expansion of CAG repeats on Chromosome 4
- c. Missense mutation in p-App gene
- d. Mutation in the APOE4 allele
- e. X-linked recessive trait

**Correct answer:** B

Huntington’s Disease is a classic trinucleotide repeat disorder (B), where there is an expansion of CAG repeats on Chromosome 4. Huntington’s is inherited autosomally (not X-linked [E]) and involves atrophy of cholinergic and GABAergic neurons in the caudate.

HOX genes (A) are responsible for general anterior-posterior patterning of the nervous system; disruption of proper HOX regulation would result in developmental disorders.

The APOE4 allele (C) and p-App (D) genes are both associated with the familial form of Alzheimer’s disease.
A 24-year-old man with a 0.5 pack year smoking history presents with progressive dyspnea and cough. Pulmonary function tests show that he has prolonged expiratory time. Genetic analysis reveals that he has alpha1-antitrypsin deficiency. What type of emphysema will this man develop?

a. Bullous emphysema  
b. Centrilobular emphysema  
c. Irregular emphysema  
d. Panacinar emphysema  
e. Paraseptal emphysema

**Correct answer:** D

Alpha1-antitrypsin deficiency is a hereditary disorder that results in defective secretion of alpha1-antitrypsin deficiency by the liver. In homozygotes, this eventually results in panacinar emphysema (choice D) and hepatic cirrhosis by young adulthood.

Bullous emphysema (choice A), centrilobular emphysema (choice B), irregular emphysema (choice C), and paraseptal emphysema (choice E) are other patterns of emphysema not associated with alpha1-antitrypsin deficiency.
An 13-year-old girl presents for evaluation of short stature (<10th percentile) and absence of breast development. Her mother reports that the patient was the same height as her peers until a few years ago. On physical exam, patient is found to have appropriate pubic hair and normal external female genitalia. There were no other significant findings except for elevated blood pressure in both arms and weak femoral pulses. What is the patient's most likely cardiac defect?

a. Tetralogy of Fallot
b. Epstein's anomaly
c. Patent ductus arteriosus
d. Coarctation of the aorta
e. Eisenmenger's syndrome

**Correct answer:** D

The patient has Turner's syndrome and thus most likely has coarctation of the aorta. Turner's syndrome is associated with type I coarctation, which involves narrowing of the aorta distal to the left subclavian but proximal to the insertion of the ductus arteriosus or preductal.

Tetralogy of Fallot includes VSD, pulmonary stenosis, right ventricular hypertrophy, and overriding aorta. This defect is associated with cyanosis and would be diagnosed at a younger age.

Epstein's anomaly (found in babies of mothers taking lithium in the 1st trimester) is a downward displaced tricuspid valve. This type of congenital heart defect is not associated with cyanosis (similar to coarctation of the aorta), but it is not associated with Turner's syndrome.

A patent ductus arteriosus is associated with rubella and prematurity, not Turner's syndrome. A PDA often has a continuous, machine-like murmur. In addition, this defect is associated with the later development of cyanosis. First, there is a left-to-right shunt that then develops into the right to left shunt of Eisenmenger's.

Eisenmenger's syndrome is the right to left shunt that develops after an initial left-to-right shunt. It is associated with thickening of the pulmonary artery walls and the common symptoms include cyanosis, clubbing, pulmonary hypertension, and heart failure. The congenital defects that may result in Eisenmenger's if they are not corrected include VSD, ASD, PDA, persistent truncus arteriosus, and anomalous pulmonary venous drainage. Eisenmenger's is not associated with coarctation of the aorta or with Turner's syndrome.
A 27-year-old man is involved in a motor vehicle accident and sustains massive blood loss. Paramedics report an initial blood pressure of 110/65 mmHg.

What anatomical structures are involved in the afferent pathway of the physiologic response to maintain this patient’s blood pressure?

a. Aortic arch transmits via vagus nerve to medulla
b. Aortic body transmits via vagus nerve to medulla
c. Carotid body transmits via glossopharyngeal nerve to medulla
d. Carotid sinus transmits via glossopharyngeal nerve to medulla
e. Carotid sinus transmits via vagus nerve to medulla

**Correct answer:** D

Carotid sinus via glossopharyngeal nerve to medulla (Choice D) is correct. This baroreceptor responds to both increases and decreases in blood pressure.

Aortic arch transmits via vagus nerve to medulla (Choice A) describes an afferent pathway for detection of increased blood pressure; however, this baroreceptor does not respond to decreased blood pressure.

Aortic body transmits via vagus nerve to medulla (Choice B) is incorrect because the aortic body is a chemoreceptor, not a baroreceptor. It will respond to a decreased PO2 (less than 60 mmHg), increased PCO2, or decreased blood pH.

Carotid body transmits via glossopharyngeal nerve to medulla (Choice C) is incorrect because the carotid body is a chemoreceptor, not a baroreceptor. It will respond to a decreased PO2 (less than 60 mmHg), increased PCO2, or decreased blood pH.

Carotid sinus transmits via vagus nerve to medulla (Choice E) is incorrect because the carotid sinus baroreceptor transmits via the glossopharyngeal nerve, not vagus.
A 45-year-old man presents to his primary care physician complaining of occasional episodes of palpitations, sweating and headaches which resolve spontaneously. He reports his father had a tumor removed from the abdomen but does not know which kind. He is found to have a blood pressure of 185/100. Urine catecholamines are found to be elevated. An abdominal CT is normal. Upon further investigation, the following image is obtained. What is the most likely cause of this patient’s problem?

a. Medullary carcinoma of the thyroid  
b. Hyperparathyroidism  
c. Neurofibroma  
d. Paraganglioma  
e. Non-Hodgkin’s lymphoma  

_correct answer_: D

Pheochromocytomas are catecholamine-secreting tumors of chromaffin cells typically arising from the adrenal medulla. However, 10-15% of these tumors can be extramedullary and are usually found in the para-aortic sympathetic ganglions (termed paragangliomas). Most of them are located in the abdominal aorta, the aortic bifurcation as well as the bladder wall and carotid bifurcation. It is helpful to remember the “rule of 10” for pheos: 10% are extramedullary, 10% are bilateral, 10% are malignant.

Parathyroid and thyroid tumors, neurofibromas and non-Hodgkin’s lymphomas (Options a, b, c, e) could also be found in this region of the neck, but it is important to note that in a hypertensive, the triad of headaches, palpitations and diaphoresis carries a high sensitivity (91%) and specificity (94%) for pheochromocytoma. Furthermore, this patient had positive urine catecholamines and possibly a family history of the disease.
A 35 year-old-female presents to her gynecologist after missing her period for the past 3 months. She is sexually active and uses condoms for contraception but has not had a sexual partner in the last year. She also reports that her breasts have been leaking milk for some time. Physical examination is normal. A head MRI is shown. Which of the following structures would most likely be jeopardized if the tumor continues to increase in size?

- a. Optic chiasm
- b. Fourth ventricle
- c. Cerebral aqueduct
- d. Thalamus
- e. Caudate nucleus

**Correct answer:** A

Prolactinomas are prolactin-secreting tumors of the anterior pituitary, classically presenting with galactorrhea and amenorrhea or infertility in pre-menopausal women. Amenorrhea occurs due to prolactin’s inhibition of LH, as well as GnRH pulses. Downregulation of GnRH leads to a decrease in FSH, and subsequently, estrogen levels leading to anovulation. Fortunately, these effects lead to their early detection in pre-menopausal women compared to their male and post-menopausal counterparts. In the latter populations these tumors tend to be of greater size and do not come into clinical attention until they are large enough to affect adjacent structures. The optic chiasm (Option A) is located directly above the pituitary gland and can be compressed leading to blindness. None of the remaining options are near the pituitary gland and therefore are less likely to be affected by these tumors.
A 27 year-old woman is brought to the emergency room by ambulance after a car accident. She is complaining of a severe headache and is somewhat disoriented. Her heart rate is 110 and her blood pressure is 134/72. She has several abrasions of her face with some swelling and ecchymosis. She has a dark ecchymosis over her right mastoid process. CT scan of the head shows an intracranial hemorrhage that does not cross suture lines. The anatomic structure that passes through which of the following foramina has most likely been damaged?

a. Carotid canal  
b. Foramen magnum  
c. Foramen rotundum  
d. Foramen spinosum  
e. Optic canal  

**Correct answer:** D

This patient has a basilar skull fracture secondary to trauma sustained in a car accident. Battle's sign, ecchymosis over the mastoid process, is consistent with a basilar skull fracture. Also, this patient has an epidural hemorrhage as seen on CT scan (does not cross suture lines). This scenario is due to rupture of the middle meningeal artery, which passes through the foramen spinosum. The maxillary nerve (V2) passes through the foramen rotundum. The optic nerve and ophthalmic artery passes through the optic canal. The internal carotid artery passes through the carotid canal. The brainstem (specifically, the medulla), meninges, vertebral arteries, and the spinal roots of the accessory nerve all pass through the foramen magnum.
A 5-year-old boy is brought in by his mother because she is concerned that he might have a mass in his lower abdomen. She says it is usually noticeable, but sometimes completely goes away. On exam, the boy's intestines are palpated protruding through the internal inguinal ring. The protrusion increases with maneuvers that increase intra-abdominal pressure, but is reducible. In surgery, which of the following findings would confirm the likely diagnosis?

- a. Hernia sac following the path of the inferior epigastric artery
- b. Hernia sac inferior to the origin of inferior epigastric artery
- c. Hernia sac lateral to the inferior epigastric artery
- d. Hernia sac medial to the inferior epigastric artery
- e. Hernia sac posterior (deep) to the inferior epigastric artery

**Correct answer:** C

This boy has an indirect inguinal hernia. It is caused by the failure of closure of the processus vaginalis (i.e. a 'patent' processus vaginalis), which allows intra-abdominal contents to enter the inguinal canal. Direct inguinal hernias occur in older patients, and are characterized by a bulge medial to the inferior epigastric artery.
A 12-year-old boy is brought to the physician by his father because of redness and swelling of his left foot for 24 hours. Three days ago, the boy scraped his foot while wading in a drainage ditch. Examination of the left foot shows a purulent abrasion with edema, erythema, and tenderness on the lateral side. Infection is most likely to next spread from the lateral side of the foot to the regional lymph nodes in which of the following areas?

- a. Lateral surface of the thigh
- b. Medial malleolus, posteriorly
- c. Popliteal fossa
- d. Sole of the foot
- e. Superficial inguinal area

**Correct answer:** C

The boy's wound appears to be superficial - he "scraped" his foot which is now infected (likely by a Gram positive organism such as Strep A or Staph). The superficial lymphatic drainage of the lower extremity roughly follows the superficial vessels. In this case of the lateral foot, the lymphatics drain along the path of the small saphenous vein to the popliteal fossa.
A 25-year-old woman is undergoing a nephrectomy in order to donate her kidney to her older sister, who has renal failure. The surgeon accidentally cuts a vein draining into the inferior aspect of the renal vein on the side of the kidney which is to be removed. What is the most likely identity of this vein?

a. Left ovarian vein  
b. Left suprarenal vein  
c. Right ovarian vein  
d. Right suprarenal vein  
e. Splenic vein

**correct answer:** A

The left ovarian vein (or testicular vein in men) enters the inferior aspect of the left renal vein.

The left suprarenal vein (B) also enters the left renal vein, but at its superior aspect.

The right ovarian (C) and right suprarenal (D) veins drain directly into the inferior vena cava.

The splenic vein (E) joins with the superior mesenteric vein to form the portal vein.

Of note, in kidney transplantation, the left kidney is usually preferred because the left renal vein is longer than the right renal vein (the left renal vein has to cross over the vertebral column to get to the inferior vena cava while the right does not).
A 13-year-old figure skater botches a quadruple lutz and falls on her right outstretched hand. Several weeks later a cast is removed for a healed scaphoid fracture. She begins having episodes of intense burning pain, sweating, swelling, and redness that begin in her right hand and move up her forearm, sometimes up to her shoulder. These episodes often occur when she is angry or upset. On exam her arm shows no visible abnormalities. Which of the following nerve types are involved in the pathogenesis of her disorder?

a. General somatic afferent
b. General somatic efferent
c. General visceral afferent
d. Special somatic afferent
e. Special visceral afferent

correct answer: C

Reflex sympathetic dystrophy, aka complex regional pain syndrome, is a mysterious disorder that often occurs after injury to an extremity. It is thought to be due to formation of an abnormal sympathetic reflex arc that becomes modulated by cortical inputs in times of stress. The key to this diagnosis is that it does not obey any specific nerve distribution and symptoms reflect vasospasm. Sympathetic reflexes are autonomic, consisting of a general visceral afferent (GVA) arm and general visceral efferent (GVE) arm. Even if you did not know the diagnosis, the episodic diaphoresis and vasodilation should point you towards an autonomic pathogenesis. Special somatic afferents (SSA) are only involved in sensation of vision, hearing and balance. General somatic afferents (GSA) provide sensation from all parts of the body except the gut, smooth muscle, and glands. Special visceral afferents (SVA) sense taste and smell. General somatic efferents (GSE) innervate somatic muscles. Do you really have to remember these dreaded 3-letter abbreviations? Unfortunately yes, you are expected to recognize them quickly and know what they do.
A 17-year-old male falls on his wrist while playing basketball. After taking a history, you notice on physical exam that he has marked tenderness within the "anatomical snuff box." What is the likely diagnosis?

- a. Fracture of the hook of the hamate bone
- b. Fracture of the pisiform bone
- c. Fracture of the scaphoid bone
- d. Tendonitis
- e. Wrist sprain

**Correct answer: C**

The anatomical snuff box is the triangular space formed by the extensor pollicis longus (medial border), extensor pollicis brevis & abductor pollicis longus (lateral border), and the distal radial styloid. The trapezium and scaphoid bones form the floor or base of the snuff box. Tenderness in the anatomical snuffbox indicates scaphoid fracture until proven otherwise!
A 19-year-old construction worker sustains a superficial laceration of the anterior wrist just proximal to the thenar and hypothenar eminences. Sensation is intact. Which of these tendons has most likely been severed?

a. Extensor carpi ulnaris
b. Flexor digitorum profundus
c. Flexor digitorum superficialis
d. Flexor pollicis longus
e. Palmaris longus

correct answer: E

The most superficial of the above tendons is the palmaris longus (E). The extensor carpi ulnaris (A) is not on the anterior wrist. The flexor digitorum profundus (B), flexor digitorum superficialis (C), and flexor pollicis longus (D) tendons are all in the carpal tunnel, and are thus covered by the flexor retinaculum, whereas the palmaris longus is not.
A 67 year-old woman complains of decreasing ability to make it to the bathroom before urinating. She reports that the problem started intermittently "a few years ago," but is now almost a daily occurrence. Past obstetric history is significant for six normal vaginal deliveries. You recommend daily exercises to strengthen muscles of the urogenital diaphragm. Strengthening which of the following muscles will most help the woman's incontinence?

a. Gluteus minimus
b. Levator ani
c. Obturator internus
d. Piriformis
e. Pubococcygeus

**correct answer:** E
The pubococcygeus most directly controls urine flow and is the muscle that contracts during orgasm. It surrounds the anus, the bladder opening, and, in women, the vagina. The pubococcygeus extends from the pubic bone to the coccyx.
A man pushes a piano across the floor. At the wrist, the force is transmitted from the carpal bones to the radius. At the elbow, the force is transmitted from the ulna to the humerus. Which of the following structures transmits the force from the radius to the ulna?

a. Annular ligament  
b. Bicipital aponeurosis  
c. Flexor retinaculum  
d. Intermuscular septum  
e. Interosseous membrane

**correct answer: E**

The interosseous membrane (E) is a fibrous sheet between the radius and ulna, forming the radio-ulnar syndesmosis. It divides the forearm into anterior and posterior compartments, and transmits force from the radius to the ulna.

The annular ligament (A) comes off the lateral side of the ulna and encircles the proximal radius. It prevents translation of the proximal radius during pronation, and has no effect on axial forces like the force described here.

The bicipital aponeurosis (B), a.k.a. lacertus fibrosus, extends medially from the biceps tendon to reinforce the cubital fossa. It does not transmit force from the radius to the ulna.

The flexor retinaculum (C), a.k.a. transverse carpal ligament, forms the roof of the carpal tunnel. It does not transmit force from the radius to the ulna.

The intermuscular septum (D) is one of two (lateral or medial) structures arising from the humerus, separating the anterior and posterior compartments of the upper arm. It does not transmit force from the radius to the ulna.
A 60-year-old woman who has had four children and completed menopause 6 years ago develops urinary incontinence whenever she coughs, sneezes, or laughs. The physician should suggest exercises to strengthen which of the following muscles?

a. Detrusor
b. Obturator internus
c. Piriformis
d. Rectus abdominis
e. Urogenital diaphragm

correct answer: E

In women (especially in the elderly or those with multiple vaginal deliveries), there is tendency for the urogenital diaphragm (E) to weaken causing incontinence due to increase in the abdominal pressure (coughing, sneezing, laughing). Strengthening these muscles (sphincter urethrae muscle (external urethral muscle), deep transverse perineal muscle, superficial transverse perineal muscle and perineal membrane) can often help this incontinence.

The detrusor (A) is the muscle around the bladder that helps expel urine by squeezing the bladder.
The obturator internus (B) and the piriformis (C) stabilize the hip.
The rectus abdominis (D) is muscle of the abdominal wall.
A 40-year-old woman is diagnosed with a lesion of the facial nerve proximal to the origin of the chorda tympani in the posterior wall of the tympanic cavity. Which of the following functions would most likely remain intact in this patient?

- a. Control of muscles in lower half of face
- b. Control of muscles in upper half of face
- c. Salivary secretion by submandibular gland
- d. Taste sensation from anterior two-thirds of tongue
- e. Tear production by lacrimal gland

**Correct answer**: E

Choice E (Tear production by lacrimal gland) is correct. The greater petrosal nerve branches from the facial nerve at the level of the geniculate ganglion. It is the source of preganglionic parasympathetic nerve fibers for the lacrimal gland.

Choice A (control of muscles in lower half of face) and Choice B (control of muscles in upper half of face) are incorrect. A lesion of the facial nerve proximal to its terminal branches (the temporal, zygomatic, buccal, marginal mandibular, and cervical nerves) would result in paralysis of all ipsilateral muscles of facial expression.

Choice C (salivary secretion by submandibular gland) and Choice D (taste sensation from anterior two-thirds of tongue) are incorrect. The chorda tympani carries taste from the anterior two-thirds of the tongue and preganglionic parasympathetic nerve fibers for the submandibular gland. Therefore, a lesion superior (proximal) to its branching point would result in loss of these functions.
Following a circus accident, a man is unable to abduct his left arm from a 90 degree position to 180 degrees. Impairment of which of the following nerves could explain this finding?

a. Axillary nerve
b. Long thoracic nerve
c. Musculocutaneous nerve
d. Radial nerve
e. Thoracodorsal nerve

correct answer: B

The long thoracic nerve innervates the serratus anterior muscle, which- among its other functions- upwardly rotates the scapula allowing the arms to reach a 180 degree position. The thoracodorsal nerve innervates the latissimus dorsi, which has no function in arm abduction. The axillary nerve innervates the deltoid, which abducts the arm to 90 degrees. The radial nerve innervates the posterior muscles of the arm and forearm, which are not involved in arm abduction. The musculocutaneous nerve innervates muscles of the anterior arm, which are not involved in arm abduction.
A neuroscientist conducting experiments on the function of various hypothalamic nuclei uses an electrode to burn a small lesion in a rat’s brain. Over the next few weeks, the rat begins to eat uncontrollably and increases its mass by 60%. What hypothalamic nucleus was ablated?

a. Anterior nucleus  
b. Lateral nucleus  
c. Septate nucleus  
d. Suprachiasmatic nucleus  
e. Ventromedial nucleus

**correct answer:** E  
The ventromedial nucleus is responsible for giving a sense of satiety; its destruction would cause hyperphagia. By contrast, the lateral nucleus is responsible for the feeling of hunger.

The septate nucleus is involved in sexual functioning. The suprachiasmatic nucleus helps control the circadian cycle. The anterior nucleus helps control parasympathetic tone and to cool the body when it is hot.

Knowledge of the hypothalamic nuclei may seem esoteric, but is actually frequently tested on the USMLE.
A 60-year-old man develops a tremor in his fingers. The tremor is most pronounced when he reaches for his coffee cup or points to an object. Which of the following components of the motor system is most likely to be involved?

a. Basal ganglia
b. Cerebellar hemisphere
c. Cerebellar vermis
d. Frontal eye field
e. Motor nucleus of the thalamus

correct answer: B

The tremor described in the question stem is an intention tremor since it is most obvious as the patient’s hand moves closer to his target (i.e., his coffee cup), and is not present while at rest. Intention tremors are common in essential tremor, intoxication, multiple sclerosis. They are a sign of cerebellar dysfunction. The cerebellum is split into the vermis and hemispheres. The cerebellar hemisphere (B) regulates the coordination of the ipsilateral extremities and therefore would be involved in an intention tremor. The cerebellar vermis (C) regulates the trunk and midline structures and its dysfunction would cause ataxia (balance problems).

Basal ganglia (A) involvement would cause rest tremor like those seen in Parkinson's.

Frontal eye field (D) would cause vision problems but not a tremor.

Motor nucleus of the thalamus (E) would most likely cause strength deficits, but not a tremor.
A 33-year-old woman undergoes abdominal surgery to correct a ureteral stricture of unknown origin. Anterior to the psoas at the level of the aortic bifurcation, the surgeon observes three tubular structures: gonadal artery, gonadal vein, and ureter. In the absence of anatomic variations, which description best identifies the ureter?

a. Most anterior structure at the level described; inferiorly it courses anterior to the iliac bifurcation.
b. Most anterior structure at the level described; inferiorly it courses posterior to the iliac bifurcation.
c. Most posterior structure at the level described; inferiorly it courses anterior to the iliac bifurcation.
d. Most posterior structure at the level described; inferiorly it courses posterior to the iliac bifurcation.
e. Neither most anterior nor most posterior structure at the level described; inferiorly it courses anterior to the iliac bifurcation.

correct answer: C

At the level of the aortic bifurcation, the ureter typically runs posterior to the gonadal artery and vein. When crossing the pelvic brim at the level of the iliac bifurcation, the ureter courses anteriorly. On its way to the urinary bladder, the ureter is typically then posterior to the uterine artery (or vas deferens in men); this final relationship gives rise to the “water [urine] under the bridge” mnemonic.
A 25-year-old female patient with a history of pelvic inflammatory disease (PID) presents for preconception counseling. She heard from a friend that she is at higher risk for an ectopic pregnancy due to her history of PID, and she is curious about where fertilization and implantation usually take place. You tell her that fertilization normally occurs in which of the following anatomic locations?

- a. Ampulla of fallopian tube
- b. Infundibulum of fallopian tube
- c. Isthmus of fallopian tube
- d. Isthmus of uterus
- e. Vagina

**correct answer:** A

The correct answer is A. Fertilization normally occurs in the ampulla of the fallopian tube (Choice A), which is the longest and widest part. Less commonly, it occurs in the infundibulum of the fallopian tube (Choice B), which is the funnel-shaped termination formed of fimbriae. After fertilization, the ovum remains in the fallopian tube for approximately 72 hours before being swept down the fallopian tube by cilia into the uterine cavity, where it implants. PID is a risk factor for ectopic pregnancy, as it can lead to tubal scarring or decreased peristalsis which, in turn, can lead to abnormal implantation outside the uterine cavity. Fertilization does not normally occur in the isthmus of the fallopian tube (Choice C), isthmus of the uterus (Choice D), or the vagina (Choice E).
A 22-year-old man presents to his physician complaining of nasal congestion. Rhinoscopy demonstrates focal inflammation with mucosal edema in the inferior nasal meatus. Drainage from which of the following structures is most likely to be obstructed?

- a. Anterior ethmoidal air cells
- b. Frontonasal duct
- c. Maxillary sinus
- d. Nasolacrimal duct
- e. Posterior ethmoidal air cells

correct answer: D

Nasolacrimal duct (Choice D) is correct. Tears drain from the conjunctival sac into the lacrimal sac, through the nasolacrimal duct into the inferior nasal meatus.

Anterior ethmoidal air cells (Choice A), frontonasal duct (Choice B), and maxillary sinus (Choice C) all typically drain into the middle meatus. They should not be affected by obstruction of the inferior nasal meatus.

Posterior ethmoidal air cells (Choice E) usually drain into the superior meatus. They should not be affected by obstruction of the inferior nasal meatus.
The following is true of cardiac muscle cells:

a. Intercalated discs contain desmosomes which are directly responsible for electrochemical and metabolic coupling
b. Intercalated discs contain gap junctions which are directly responsible for the spread of action potentials between cells
c. Intercalated discs are dense regions where calcium accumulates after it is released from the sarcoplasmic reticulum
d. The arrival of an action potential leads to the relaxation of the muscle cell
e. The arrival of an action potential allows for the sequestration of calcium in the sarcoplasmic reticulum

**Correct answer:** B

Gap junctions are key components of the intercalated discs in cardiac muscle cells. They allow for direct ion flow between the cells, leading to the spread of APs and synchronous muscle contraction.
A 45 year-old man with a history of alcoholism develops pancreatitis. What pathologic changes would likely be seen in this patient’s pancreas?

- a. Eosinophilic deposits in vascular walls
- b. Granulomatous inflammation with some preservation of tissue architecture
- c. Liquefied necrotic tissue
- d. Necrotic fat cells, calcium soap formation, and lipid-laden macrophages
- e. Tissue architecture well-preserved, with nuclear changes

**Correct answer: D**

Enzymatic fat necrosis is seen in pancreatitis, and is characterized histologically by necrotic fat cells, calcium soap formation, and lipid-laden macrophages. The two most frequent causes of pancreatitis are gall stones and alcohol.

Eosinophilic deposits in vascular walls is representative of fibrinoid necrosis. Granulomatous inflammation with some preservation of tissue architecture is representative of caseous necrosis. Liquefied necrotic tissue is representative of liquefactive necrosis. Tissue architecture well-preserved, with nuclear changes is representative of coagulative necrosis.
A 46 year-old white male with a family history of esophageal adenocarcinoma presents complaining of heartburn. On endoscopy, a crescent-shaped area of erythematous mucosa is seen that extends above the gastroesophageal junction. A biopsy of this area of mucosa is taken. What histological features will most likely be seen on H&E stain?

- Columnar epithelium with goblet cells
- Columnar epithelium with parietal cells
- Keratinized squamous epithelium
- Non-keratinized squamous epithelium
- Transitional epithelium

**Correct answer:** A

This erythematous mucosa is likely Barrett's esophagus, or metaplasia of the normal squamous epithelium of the distal esophagus to columnar epithelium. This usually occurs in the setting of gastroesophageal reflux. Barrett's is considered pre-malignant for esophageal adenocarcinoma, but not for squamous cell carcinoma. Goblet cells are mucous-secreting cells that will easily be seen in Barrett’s on H&E stain. Parietal cells are not associated with the development of Barrett's – furthermore, they would not easily be seen on H&E stain. Transitional epithelium is seen in the urinary tract.
A pathologist receives a normal specimen resected from a patient’s gastrointestinal tract. Under the microscope, she sees that the mucosa contains columnar cells lacking villi. There are many straight and long tubular glands. This specimen most likely originates from which portion of the GI tract?

- Esophagus
- Large intestine
- Pharynx
- Small Intestine
- Stomach

**Correct answer:** B

The large intestine (choice B) contains the cecum and the colon. Its mucosa is composed of columnar cells lacking villi, and contains openings which give way to tubular glands known as the crypts of Lieberkuhn.

The mucosa of the esophagus (choice A) is composed of thick, stratified, squamous, nonkeratinized epithelium.

The pharynx (choice C) contains stratified, squamous epithelium, as well as ciliated, pseudostratified, columnar epithelium.

Intestinal villi are the hallmark of the small intestine (choice D). They are composed of outgrowths of the mucous membrane into the lumen. Crypts of Lieberkuhn are also found in the small intestine.

The stomach (choice E) mucosa contain simple, columnar cells, but lacks the crypts of Lieberkuhn.
A 65 year-old man with no past medical history presents for his annual check-up. He notes some increased dyspnea on exertion along with an overall lethargic feeling for the past few months. Physical exam is normal with the exception of a heme-occult positive stool test, and he is subsequently found to have elevated liver function tests. Which of the following is true?

a. A peripheral blood smear will show microcytic erythrocytes
b. A CEA level will be at least twice the upper limit of normal
c. A peripheral blood smear would show polymorphonuclear cells with more than five lobes of the nucleus
d. A biopsy of the primary lesion will show signet-ring cells
e. A CEA level will be undetectable

**correct answer:** A

The answer is A. The patient most likely has adenocarcinoma of the colon until proven otherwise. The heme-occult stool test has >95% sensitivity for this disease and colon cancer also can lead to iron-deficiency anemia. Iron-deficiency anemia manifests as microcytic hypochromic erythrocytes on a peripheral blood smear.

Choice B is incorrect; while a CEA level may be monitored prior to resection and afterwards to detect recurrence or liver metastases, it may not be elevated in many patients with colorectal cancer. CEA levels should not be used to diagnose or screen for colorectal cancer.

Choice C is incorrect; hypersegmented PMNs are the first indication of B12 or folate deficiency anemias.

Choice D is incorrect; a biopsy of adenocarcinoma of the colon will most likely not show signet rings. This pathology finding is more likely to be associated with gastric cancer although it is sometimes found in colon cancer.

Choice E is incorrect; CEA levels are rarely undetectable. Also see explanation for choice B above.
A 20-year-old female sees her physician for diarrhea and fatigue with a 20 pound weight loss over the past 6 months. On exam, she is afebrile and has mild muscle wasting, but her strength is normal. Stool studies do not reveal blood, ova, or parasites. A biopsy of the jejunum is taken and microscopically reviewed. The patient is placed on a special diet with no wheat or rye products. The change in diet produces a dramatic improvement. Which of the following microscopic features is most likely to be seen in the biopsy?

a. Crypt abscesses
b. Foamy macrophages within the lamina propria
c. Lymphatic obstruction
d. Noncaseating granulomas
e. Villous blunting and flattening

**Correct answer:** E

This patient has malabsorption that responded to dietary treatment; likely celiac disease (gluten sensitivity). The histological features of celiac disease are flattening of the mucosa, diffuse and severe atrophy of the villi, and chronic inflammation of the lamina propria.

Crypt abscesses (choice A) is incorrect; these are nonspecific and can be seen in inflammatory bowel disease.

Foamy macrophages (choice B) and lymphatic obstruction (choice C) are incorrect; these features are seen in Whipple’s disease. PAS-positive granules can show Tropheryma whippelii under electron microscope.

Noncaseating granulomas (choice D) is incorrect; these are found in Crohn’s disease.
A 34 year old woman presents to her general physician with nonspecific fever of five days’ duration. She says that there are swollen “bumps” in her right axillary region which are painful. She complains of fatigue, low-grade fever, and some stomach pain. Physical exam reveals tender right axillary lymphadenopathy. She also has a minor laceration on her right forearm that her cat gave her a week ago. She admits to a 5 kg weight loss, but says she is not hungry. What findings are likely to be prominent if a biopsy was performed of a right axillary lymph node?

a. Prominent germinal centers
b. Effacement of the nodal architecture
c. Proliferation of cytotoxic CD8 T cells
d. Atypical lymphocytes
e. Numerous band cells

correct answer: A

This patient is most likely suffering from "cat-scratch disease", a bacterial infection caused by Bartonella henselae. This unilateral lymphadenopathy on the side of the scratch combined with nonspecific constitutional symptoms is characteristic of the disease. The patient thus likely has expansion of germinal centers, which are aggregates of B cells that can occur upon antigen presentation.

Effacement of nodal architecture is characteristic of lymphoma, especially Hodgkin lymphoma in a female patient of this age.

Proliferation of cytotoxic CD8 T cells can occur during a delayed hypersensitivity reaction to antigens such as tuberculosis.

Atypical lymphocytes are characteristic to infection with Ebstein-Barr Virus or cytomegalovirus.

Numerous band cells are more common in acute septic infections such as those by pneumococcus.
A 41-year-old woman with no significant medical history presents complaining of fatigue and lightheadedness. Physical exam is notable for conjunctival pallor, bilateral brisk patellar reflexes, slightly ataxic gait, and impaired position and vibration sense in the feet bilaterally. Her hematocrit is 23%. Serologic studies are positive for anti-parietal cell antibodies. What would you expect to see on peripheral blood smear?
   a. Burr cells
   b. Hypersegmented neutrophils
   c. Howell-Jolly bodies
   d. Schistocytes
   e. Spur cells

**correct answer:** B

This patient has pernicious anemia, autoimmune attack on parietal cells causing decreased production of intrinsic factor and resultant vitamin B12 deficiency. B12 deficiency causes a megaloblastic anemia (with hypersegmented neutrophils on blood smear), stomatitis, glossitis, and subacute combined degeneration of peripheral nerves and spinal cord (which can cause ataxia, hyperreflexia, and impaired position/vibration sense). Burr cells (A) and spur cells (E) are types of poikilocytes - burr cells suggest renal disease and spur cells (acanthocytes) suggest liver disease or lipid abnormalities. Howell-Jolly bodies (C) are clusters of DNA within RBC cytoplasm indicative of asplenia or hyposplenia. Schistocytes (D) are fragmented red blood cells that are seen in microangiopathic diseases such as disseminated intravascular coagulation (DIC), thrombotic thrombocytopenic purpura (TTP), and hemolytic uremic syndrome (HUS).
In young infants as well as geriatrics patients, "bandemia" (increased band cells) is a sign of acute bacterial infection. When a band cells differentiates, which of the following types of cells is a possible mature cell type it can become?

a. Erythrocyte  
b. Lymphocyte  
c. Neutrophil  
d. Monocyte  
e. Platelet

**correct answer:** C

Myeloblasts differentiate into promyelocytes, myelocytes, metamyelocytes, then band cells, and finally branch into neutrophils, eosinophils and basophils. These types of cells are all known as granulocytes. Lymphoblasts differentiate into B cell and T cell lymphocytes, as well as natural killer cells. Monoblasts differentiate into monocytes. Megakaryoblasts turn into megakaryocytes, which produce platelets.
A physician-scientist is conducting an experiment in which she tries to stimulate growth of cartilage in vitro. She wishes to use a substance to stimulate histogenesis. Which substance would be best suited to her purposes based on its stimulation of cartilage growth in vivo?

- a. Cortisone
- b. Estrogen
- c. Thyroxine
- d. Vitamin A
- e. Vitamin D

**Correct answer:** C

Cartilage growth is stimulated by thyroxine (Choice C), testosterone and growth hormone.

It is inhibited by estrogen (Choice B) and cortisone (Choice A).

Vitamin A (Choice D) and Vitamin D (Choice E) are involved in bone mineralization.
A 52-year-old premenopausal African-American woman reports excessive bleeding from her vagina over the last 7 days. Imaging of the uterus shows a single bulky, oval mass originating deep to the endometrium. Biopsy reveals areas of necrosis and hemorrhage, with a high degree of cellular atypia and mitotic index. It is characterized as non-estrogen sensitive and appears to have arisen de novo. Which of the following is the best classification for such a tumor?

a. Endometrial carcinoma  
b. Germinoma  
c. Leiomyosarcoma  
d. Leiomyoma  
e. Ovarian cyst

**Correct answer: C**

Leiomyosarcomas (C) are bulky and aggressive tumors arising from the uterine myometrium. The malignant characteristics with areas of necrosis and hemorrhage should identify this as distinct from leiomyomas, benign tumors that typically do not progress to malignancy. In general, this tumor has a very poor prognosis.

Endometrial carcinoma (A) are the most common gynecologic malignancy. However, these tumors are usually preceded by endometrial hyperplasia, and by definition originate in the endometrium (not deep to it, as in this case). The most common presenting symptom is postmenopausal bleeding.

In females, germinomas (B) and cystadenomas (E) present on the ovaries, and not in the uterus, as described here.

While leiomyomas (D) are the most common female tumors, malignant transformation as seen here is rare. These benign 'fibroid' tumors do not progress into leiomyosarcomas, and typically display estrogen sensitivity, unlike the lesion seen here.
An infant is born prematurely at 32 weeks in respiratory distress. A chest X-ray shows signs of neonatal respiratory distress syndrome (RDS) and the infant is treated with exogenous surfactant. Which of the following types of pulmonary cells makes surfactant?

a. Type I alveolar cells  
b. Type II alveolar cells  
c. Capillary endothelial cells  
d. Clara cells  
e. Pseudocolumnar ciliated cells

**Correct answer:** B

Surfactant is made by type II alveolar cells and stored in lamellar bodies within these cells. They start developing after 24 weeks gestation, which is one of the major reasons birth before 24 weeks is not compatible with life. Type I alveolar cells make up the majority of the alveolar surface. Capillary endothelial cells secrete angiotensin-converting enzyme (ACE). Clara cells are mostly found in the primary bronchioles and contain P450 enzymes to detoxify harmful chemicals in the lungs. Pseudocolumnar ciliated cells line the trachea, creating a "muco-ciliary escalator" to expel toxins/dust in the upper airways.
A researcher is investigating the distribution of goblet cells in the respiratory system. She injects dye that will be picked up by goblet cells into an animal model. Where is the dye most likely to be absent?

- a. Intrapulmonary bronchi
- b. Larynx
- c. Nasopharynx
- d. Terminal bronchioles
- e. Trachea

correct answer: D

Goblet cells are glandular cells that secrete mucous. They are found in many places in the respiratory system, including the Nasopharynx (Choice C), Larynx (Choice B), Intrapulmonary bronchi (choice A) and Trachea (choice E), but are not found in the terminal bronchioles (Choice D).
A 71-year-old female with no significant past medical history presents to your office complaining that her friends have noticed a slight yellowing of her skin over the past month. She denies any abdominal pain, but reports a recent 15-lb weight loss as well as persistent "itching all over." On physical exam, you palpate a large, round cystic mass in right upper quadrant which you think may be her gallbladder. Which of the following serum markers is likely to be abnormally elevated given the most likely diagnosis?

- a. CA-125
- b. CEA
- c. CA-19-9
- d. AFP
- e. Beta HCG

**Correct answer:** C

Painless jaundice is the classic presentation of pancreatic cancer. A mass at the head of the pancreas blocks the common bile duct causing an obstructive jaundice. Bile salts accumulate in the dermis causing marked pruritis. The gallbladder may become distended and palpable on physical exam (Courvoisier's sign). The weight loss is additional sign of malignancy.

CA-19-9 is the tumor marker most closely associated with pancreatic cancer.

CA-125 is associated with ovarian cancer.

CEA is associated with colon cancer and other GI malignancies.

AFP is associated with hepatocellular carcinoma as well as yolk sac carcinoma (a type of testicular nonseminoma).

Beta HCG is associated with choriocarcinoma (another type of testicular nonseminoma).
A 28 year-old male of Jewish descent presents with abdominal pain, diarrhea and fever. Physical exam reveals a tender abdomen and a perianal fistula. A biopsy reveals chronic inflammation in the small intestine involving all layers of the intestinal wall. This patient may be treated with a monoclonal antibody which blocks which of the following cytokines?

- a. IL-3
- b. IL-5
- c. IL-8
- d. IFN-Beta
- e. TNF-Alpha

**Correct answer:** E

This is a classical presentation of Crohn's disease. While the etiology of this disorder is still unknown, TNF-Alpha seems to play a key role. Blocking TNF-Alpha with a monoclonal antibody (such as Infliximab or Adalimumab) causes disease remission in a large number of Crohn's patients.

IL-3 is secreted by activated T cells and stimulates bone marrow stem cells.

IL-5 is secreted by helper T cells and promotes IgA synthesis by B cells.

IL-8 is a chemotactic factor for neutrophils.

IFN-Beta is produced by APCs and epithelial cells in response to viral infections.
A researcher performs immunohistochemical analysis on a blood sample from a patient. The researcher notices a large number of cells which stain for CD19, CD20, and IgM on their cell surface. What kind of cells are these?

a. B cells  
b. Cytotoxic T cells  
c. Helper T cells  
d. Macrophages  
e. NK cells

**Correct answer:** A

While most cells express a wide variety of proteins on their cell surface, the ones commonly used to identify cells of the immune system are as follows:

- **Helper T cells:** CD4, TCR, CD3, CD28  
- **Cytotoxic T cells:** CD8, TCR, CD3  
- **B cells:** IgM or IgD, CD19, CD20  
- **Plasma cells:** IgG  
- **NK cells:** CD16, CD56, receptors for MHC I  
- **Macrophages:** MHC II, CD14  
- **Antigen Presenting Cells (APCs):** MHC II

When studying for the exam, only the most glaring markers are important. For instance, IgM should have immediately signaled B-cell (even if you did know about CD19 and CD20). So don't bother memorizing all of the other markers... just know the fundamental ones.
A 3-year-old boy has a history of repeated pyogenic infections. He had normal antibody responses following childhood immunizations and normal recovery from chickenpox and measles. Decreased numbers or functional defects in which of the following cells best explains the cause of his infections?

a. B lymphocytes  
b. Eosinophils  
c. Macrophages  
d. Neutrophils  
e. T lymphocytes

**Correct answer:** D
The question stem provides 3 useful hints which all point to Neutrophils (D) as the answer.

First, the boy's infections are of pyogenic origin, indicating bacterial infection that results in pus formation. Pus consists primarily of expended neutrophils. Since there is a defect in the pyogenic response (resulting in repeated infection), there is most likely a defect in the action of Neutrophils.

Second, his normal antibody response to immunizations can help you rule out B Lymphocytes (A) or Macrophages (C, the most common antigen presenting cell type) as the answers.

Third, a normal recovery from chickenpox and measles (both viruses) requires intact function of T lymphocytes (E) and Eosinophils (B, because they contain abundant RNAses).

The other answers do not play a direct role in the body's response to pyogenic infections.
A full term, normal appearing baby boy is born to a 27 year old mother after a normal spontaneous vaginal delivery. During the third hour of life, the baby experiences diffuse involuntary contractures of his muscles. Physical exam reveals an otherwise normal appearing neonate, at an appropriate height and weight for gestational age. Laboratory studies reveal a serum calcium of 6.1 and a phosphorus of 3.7. During the first year of life, the neonate experiences recurrent pneumonias and upper respiratory tract infections, several of which require hospitalization. Which of the following is the mechanism most likely responsible for the clinical features seen in this child?

a. Lack of adenosine deaminase (ADA)
b. Failure of differentiation of pre-B cells into mature B-cells
c. Transient immunoglobulin deficiency
d. Failure of differentiation of B cells into plasma cells
e. Malformation of the third and fourth pharyngeal pouches

**correct answer:** E

The stem describes an infant with DiGeorge syndrome secondary to malformation of the third and fourth pharyngeal pouches (option E), most being secondary to a 22q11.2 deletion. Common features seen in DiGeorge syndrome, otherwise known as velocardiofacial syndrome, can be easily remembered by the pneumonic "CATCH 22" -- Cardiac defects, Abnormal facial features, Thymic aplasia, Cleft palate, Hypocalcemia, and chromosome 22. Due to the thymic aplasia, these patients are T-cell deficient, predisposing them to recurrent fungal, viral and protozoal infections, as is described in this patient. The clue in this question stem is the involuntary muscle contractures (tetany) the infant experienced soon after birth. This is to clue the reader into the hypocalcemia (secondary to absence of the parathyroid gland), a distinguishing feature seen in DiGeorge's syndrome.

Option A- lack of adenosine deaminase, is a notable cause of the majority of cases of severe combined immunodeficiency (SCID). Option B- failure of B cell maturation- is the mechanism behind Bruton's agammaglobulinemia, an X linked recessive disorder seen exclusively in males. Option C- describes the transient hypogammaglobulinemia seen in normal infants around 5-6 months of age as levels of maternal IgG decline. Option D- failure of B cell maturation into plasma cells, is the mechanism behind common variable immunodeficiency (CVI).
A 48-year-old man receives a liver transplant for end-stage alcoholic cirrhosis. Three weeks after the transplant surgery, the man experiences fevers, chills and fatigue related to an acute rejection of the transplant. The host cells involved in this response recognize foreign antigens presented by antigen presenting cells (APC). These antigens are presented to host cells on which of the following APC receptor types?

a. B7  
b. CD28  
c. IL-2  
d. MHC Class I  
e. MHC Class II

**Correct answer:** E

Acute transplant rejection is a cytotoxic T-cell mediated process whereby cytotoxic T-cells (Tc) are indirectly activated by helper T cells (Th). Th possess a T-cell receptor (TCR) that interacts with MHC Class II antigen presenting receptors on APCs. B7 and CD28 costimulation is necessary for Th activation, but does not involve actual presentation of the antigen. IL-2, a cytokine secreted by activated Th cells, serves to stimulate growth of Th and Tc's.
A 24-year-old man with no past medical history develops urticaria and angioedema within 2 hours of taking aspirin for a headache. It is the first time he has taken aspirin in years, but he does not recall having an adverse reaction to it in the past. What kind of hypersensitivity reaction does this represent?

a. Cell-mediated  
b. Cytotoxic  
c. IgA-mediated  
d. IgE-mediated  
e. Immune complex-mediated

**Correct answer:** D

This represents an IgE-mediated (also called anaphylactic or type I) hypersensitivity reaction, where mast cells release histamine and other substances that result in vasodilation and increased vascular permeability, resulting in urticaria (hives) and angioedema (the same mechanism as urticaria, but affecting subcutaneous tissue).
An emergency-room patient is in acute distress. She is hyperventilating and feverish. Her pulse rate is 100 and her blood pressure is 90/50. A blood culture reveals high levels of gram-negative bacteria. Her condition is mediated by the systemic effects of which of the following cytokines?

a. IFN-Beta  
b. IL-10  
c. IL-2  
d. IL-3  
e. TNF-Alpha

**correct answer:** E

This patient displays a classical case of septic shock, of which TNF-Alpha is a key mediator. Toll-like receptors on the surface of macrophages bind bacterial lipopolysaccharide and activate a signaling cascade which causes the macrophages to release TNF-Alpha and other cytokines (IL-1, IL-6, and IL-8.) Infection in the bloodstream (sepsis) causes macrophages in the liver, spleen and blood to release large quantities of TNF-Alpha. The vasodilation and increased permeability of blood vessels induced by the TNF-Alpha leads to rapid loss of plasma volume. TNF-Alpha can also cause disseminated intravascular coagulation. These conditions cause septic shock, which may result in hypoperfusion of vital organs, leading to multi-organ failure.

The other cytokines in the answer choices do not cause septic shock.

Reference:  
Janeway Immunobiology, 5th Ed.
Flow cytometric data of T lymphocytes stained with fluorescent antibody to CD4+ and CD8+ antigens from a healthy person are shown in the figure. The lymphocytes from which of the following quadrants will respond most vigorously to immunization with hepatitis A virus vaccine?

a. Quadrant A  
b. Quadrant B  
c. Quadrant C  
d. Quadrant D  
e. N/A

**Correct answer:** D  
Quadrant D (D) contains CD4+ T lymphocytes (a.k.a “helper” T cells). This population is activated by antigen presented on major histocompatibility complex (MHC) class II molecules, which are only expressed on “professional” antigen presenting cells (APCs). These antigens are fragments of nonself peptides internalized by APCs. This CD4+ population would respond to hepatitis A virus vaccine.

Quadrant A (A) contains CD8+ T lymphocytes (a.k.a. “cytotoxic” T cells). This population is activated by antigen presented on major histocompatibility complex (MHC) class I molecules, which are expressed on nearly all nucleated cells. Such antigens are selected at random from cytosol content, and may be self- or nonself-peptides (e.g. infectious virion). This CD8+ population would respond to virulent hepatitis A virus, but not the vaccine.

Quadrant B (B) contains CD4+ CD8+ T lymphocytes, corresponding to an intrathymic (immature) phenotype. Such cells are usually not seen peripherally, and do not express mature T cell function.

Quadrant C (C) would contain CD4- CD8- T lymphocytes. Such cells would be abnormal and would not be involved in a vaccine response.
A healthy 19-year-old man receives a tetanus immunization booster prior to induction into the US Marines. Six hours later, he has pain and massive swelling at the site of injection. The following day, the skin breaks down, forming an ulcer at the site. Which of the following events plays a critical role in this reaction?

a. Accumulation of mononuclear cells at the site of antigen injection  
b. Antigen capture by Langerhans cells in the epidermis  
c. Local fixation of complement by preformed circulating antibodies  
d. Local release of histamine  
e. Predominant synthesis of IgM antibodies

correct answer: C

This patient is having an immune complex (type III) hypersensitivity reaction to the tetanus booster commonly referred to as an Arthus reaction. Because the patient has been previously immunized, preformed circulating antibodies locally fixate complement at the injection site due to the high load of tetanus toxoid in the booster. This results in a local inflammatory vasculitis with pain, edema and skin ulceration.

Accumulation of mononuclear cells occurs in cell-mediated (type IV) hypersensitivity. Langerhans cells are dendritic cells of the skin that present foreign antigens in response to infection or foreign bodies, but antigen capture in the epidermis has no role in the Arthus reaction. Histamine release is a key component of immediate (type I) hypersensitivity. IgM and IgG proliferation occur in antibody-mediated (type II) hypersensitivity reactions.
In the Haemophilus influenzae type b vaccine, the bacterial capsular polysaccharide is conjugated to a carrier protein to improve immunogenicity in children. Which of the following cells become activated following contact with peptides processed from this vaccine in the context of MHC type II proteins?

a. B lymphocytes
b. Macrophages
c. Natural killer cells
d. Plasma cells
e. T lymphocytes

**Correct answer:** E

Logically, a vaccine needs to work by inducing memory cells. There are two types of memory cells - T and B. Memory B cells (a variant of plasma cells) can only be produced after signaling from activated T cells. Thus, the vaccine will need to lead to the activation of T cells.

The carrier protein in the vaccine enables it to be taken up and processed by antigen-presenting cells (dendritic cells, B cells, macrophages). The peptides are presented on MHC type II, which contact and activate T-lymphocytes, starting the above immune cascade.
A 36-year-old woman with end stage renal disease secondary to polycystic kidney disease undergoes renal transplantation. On the tenth postoperative day she complains of general malaise, appears mildly ill, and has decreased urine output. What is the most likely pathologic mechanism underlying this patient’s current illness?

a. Donor T-lymphocytes reacting against recipient major histocompatibility complexes (MHCs)
b. Immune complex deposition within renal artery walls
c. Immune complex deposition within renal glomerular subendothelium
d. Preformed anti-donor antibodies reacting against donor antigens
e. Recipient cytotoxic T-lymphocytes reacting against donor major histocompatibility complexes (MHCs)

correct answer: E

Given the recent renal transplant, time course, and evidence of organ failure this patient appears to be suffering from acute transplant rejection. The most likely mechanism in acute rejection involves recipient cytotoxic T-lymphocytes reacting against donor MHCs (Choice E). Immunosuppressive medications such as cyclosporine aim to attenuate these reactions.

Donor T-lymphocytes reacting against recipient major histocompatibility complexes (MHCs) (Choice A) is the mechanism underlying graft-versus-host disease (GVHD). GVHD is typically seen in recipients of bone marrow, not solid organs, due to its high load of functional donor immune cells that may perceive recipient tissues as foreign due to MHC incompatibility. Because the recipient is immunocompromised, the donor cells cannot be destroyed or inactivated.

Immune complex deposition within renal artery walls (Choice B) is the pathologic mechanism in polyarteritis nodosa. Although polyarteritis nodosa typically involves renal and visceral vessels, the process would be more chronic than seen in this situation; moreover, renal failure would be an unusual sequela. Typical signs and symptoms include fever, weight loss, malaise, abdominal pain, melena, headache, myalgia, hypertension, neurologic dysfunction, and cutaneous eruptions.

Immune complex deposition within renal glomerular subendothelium (Choice C) is the mechanism of renal failure in systemic lupus erythematosus (SLE). Though SLE may result in renal failure, the process would be more chronic than seen in this situation. Moreover, additional stigmata of SLE would be expected, such as rash, photosensitivity, mucositis, neurologic dysfunction, pleuritis, pericarditis, hematologic dysfunction, or arthritis.

Preformed anti-donor antibodies reacting against donor antigens (Choice D) is the mechanism underlying hyperacute transplant rejection. The reaction typically occurs within minutes to hours of organ implantation. Immunosuppressive medications offer little towards reversing or attenuating these reactions.
A 6-year-old boy presents to the emergency room complaining of nausea and a new-onset rash that developed shortly after ingesting a peanut. Which of the following antibodies is most likely responsible for the type of reaction this boy is now experiencing?

- a. IgA
- b. IgD
- c. IgE
- d. IgG
- e. IgM

**Correct answer:** C

The boy is experiencing an allergic reaction after ingestion of a peanut. Dermatologic manifestations, such as urticaria, angioedema, pruritis, and erythema, are the most common effects of food allergy reactions. Gastrointestinal symptoms, such as nausea, vomiting, abdominal cramps, and diarrhea, are also common. Anaphylaxis is the most severe form of allergic reaction. In the early stages of an allergic reaction, an allergen triggers a chain reaction resulting in the production of IgE by B cells. IgE circulates in the blood and binds to Fc receptors on mast cells and basophils, resulting in their degranulation and release of inflammatory chemical mediators such as histamine, cytokines, interleukins, leukotrienes, and prostaglandins. IgE is associated with Type I hypersensitivity reactions.

IgA (Choice A) primarily functions in mucosal immunity, protecting body surfaces that are exposed to outside foreign substances.

IgD (Choice B) is found in low levels in serum where it serves an uncertain function. It is found primarily on B cell surfaces where it functions as a receptor for antigen.

IgG (Choice D) functions in the secondary antibody response and are key in fighting bacterial and viral infections. It is the only antibody that can pass through the human placenta. IgG is associated with Type II and III hypersensitivity reactions.

IgM (Choice E) functions in the primary antibody response and eliminates pathogens in the early stages of B cell-mediated immunity before there is sufficient IgG.
A 22-year-old male presents with complaints of photophobia, dysuria, and arthralgias. Upon further questioning he admits to having had a sexually transmitted disease diagnosed a few weeks ago, which was treated with antibiotics. On physical exam, his eye is erythematous with a watery discharge and his left knee is swollen, warm and tender.

**What is the most likely HLA type found in this patient?**

a. HLA B8  
b. HLA B27  
c. HLA DR2  
d. HLA DR3  
e. HLA DR5

**correct answer:** B

This patient is experiencing reactive arthritis (also known as Reiter’s Arthritis) which is a seronegative inflammatory arthritis that often follows a GU infection (like Chlamydia described in the history) or a bacterial GI infection. This group of seronegative inflammatory arthritides are associated with HLA B27. Also in this group are psoriasis, ankylosing spondylitis, and IBD.

HLA B8 is associated with Graves’ disease and celiac sprue.

HLA DR2 is associated with multiple sclerosis, SLE, and Goodpasture’s syndrome.

HLA DR3 is associated with diabetes type I (especially in conjunction with HLA DR4).

HLA DR5 is associated with pernicious anemia, and Hashimoto’s thyroiditis.
A 24-year-old female presents with monocular vision loss and a history of parasthesias and muscle weakness that has resolved. She arrives to your office with weakness in her left leg and trouble walking without assistance. Which of the following recombinant cytokines would aid in her treatment?

a. Alpha-interferon  
b. Beta-interferon  
c. Erythropoietin  
d. Gamma-interferon  
e. Granulocyte Colony Stimulating Factor (G-CSF)

**correct answer: B**

This patient presents classically with Multiple Sclerosis. Among the listed choices, Beta-interferon is specifically indicated for treatment of MS. Alpha-interferon is indicated for treatment of Hepatitis B and C, Kaposi’s Sarcoma and malignant melanoma. Erythropoietin stimulates the production of Red Blood Cells and given to combat various forms of anemia. Gamma-interferon is employed against Chronic Granulomatous Disease (NADPH oxidase deficiency). GCSF is used to stimulate bone marrow production in leukemias and during various chemotherapies.
A 10-year old child presents with a persistent sinus infection. Over the course of the last 5 years, the patient has had several dozen sinus and upper respiratory infections. Blood tests reveal abnormally low levels of IgA immunoglobulin, but normal levels of other isotypes. This patient's recurrent infections are most likely due to a defect in which of the following?

a. Fixation of complement
b. Mast cell activation
c. Mucosal immunity
d. Neutrophil activation
e. Opsonization of bacteria

**Correct answer:** C

IgA deficiency is the most common selective immunoglobulin deficiency. IgA is the main immunoglobulin isotype mediating mucosal immunity. IgA does not play a major role in fixing complement, opsonizing bacteria, mast cell activation, or neutrophil activation.
A 3-year old child has dyspnea, severe cough, and a fever. A chest X-ray and laboratory tests lead to a diagnosis of Pneumocystis jirovecii pneumonia. Further laboratory tests indicate abnormally low levels of the IgG, IgA, and IgE immunoglobulins. However, levels of IgM are far above normal. This patient’s immune deficiency is caused by a defect in which cell surface molecules?

a. CD40 ligand  
b. CD86  
c. Fas ligand  
d. TCR  
e. TNF receptor

**Correct answer:** A

This patient has hyper IgM syndrome, characterized by severe pyogenic infections starting at a young age, especially lung infections by opportunistic pathogens such as Pneumocystis jirovecii (also called Pneumocystis carinii). The molecular defect is in the CD40 ligand expressed by activated CD4+ (helper) T cells, which leads to inability to engage CD40 on B cells. Activation of CD40 is required for isotype switching of B cells; without it, B cells produce high levels of IgM but cannot switch to producing other immunoglobulin isotypes. The defect in CD40 ligand is also thought to impair activation of macrophages by CD4+ T cells, facilitating lung infections by opportunistic pathogens which are normally cleared by the alveolar macrophages.

Defects in the other cell-surface molecules in the answer choices are not typically a cause of hyper-IgM syndrome.

Reference: Janeway's Immunology, 5th Ed
A boy scout returns home after a three-day adventure in the woods with a pruritic, linear, vesicular eruption. His mother brings him to the dermatologist and he is diagnosed with poison ivy dermatitis. What type of reaction is this?

a. Type I  
b. Type II  
c. Type III  
d. Type IV  
e. Type V

Correct answer: D

Type IV hypersensitivity reactions are T-cell mediated reactions, which are responsible for the contact dermatitis seen in poison ivy. Type I reactions are anaphylactic (e.g. seasonal allergies). Type II reactions are antibody-antigen mediated (e.g. Goodpasture's nephritis). Type III reactions are immune complex mediated (e.g. rheumatoid arthritis). Type V reactions are antibody-stimulating (a subtype of Type II) (e.g. Graves disease).
A 49 year-old woman with a recent history of hypertension comes in complaining of tightening of the skin of her hands and face. BP is 142/84, and other vital signs are within normal limits. On lung exam, air entry is less than expected with deep inspiration bilaterally, with no wheezes, rales, or rhonchi. The remainder of the physical exam is within normal limits. In addition to the skin and lungs, what organ is often affected by this disease?

a. Esophagus  
b. Heart  
c. Liver  
d. Spinal cord  
e. Spleen

correct answer: A
This patient has progressive systemic sclerosis (PSS), or scleroderma. PSS can affect much of the GI tract, with the esophagus being the most common portion affected, frequently resulting in dysphagia. Findings on lung exam may represent an early finding of interstitial pulmonary fibrosis, a complication of PSS that can be fatal.
A 46-year-old man has a 4-week history of epigastric pain; test of stool is positive for occult blood. Examination of tissue obtained on biopsy of the gastric antrum shows curved bacterial rods. Which of the following additional findings is most likely?

- a. Achlorhydria
- b. Antiparietal cell antibodies
- c. Cholecystitis with antral seeding
- d. Immunodeficiency state
- e. Increased urease activity in the antrum

**Correct answer:** E

Given this patient’s presentation and the finding of curved bacterial rods, the patient is most likely infected with Helicobacter pylori. H. pylori is a major cause of peptic ulcer disease as well as gastritis, both of which can cause epigastric pain with a positive fecal occult blood test. H. pylori tests positive for oxidase, catalase, and urease. Therefore, there would most likely be increased urease activity in the antrum (choice E) of this patient.

Achlorhydria (choice A) and antiparietal cell antibodies (choice B) both describe changes seen in pernicious anemia.

Infection with H. pylori would most likely not cause cholecystitis (choice C) or an immunodeficient state (choice D).
A patient presents with severe bloody diarrhea for the last three days and right upper quadrant pain. Stool analysis reveals cysts with 4 nuclei. This patient is most likely infected with which of the following?

a. Acanthamoeba
b. Cryptosporidium
c. Entamoeba histolytica
d. Giardia lamblia
e. Leishmania donovani

**Correct answer:** C

The symptoms are a classical presentation of Entamoeba histolytica infection: bloody diarrhea (dysentery) and right upper quadrant pain. This amoeba can frequently cause liver abscesses, and can be diagnosed by observing 4-nucleated cysts in the stool. Ingested RBCs inside the amoebas and flask-shaped ulcerations in the mucosa/submucosa can also be used for diagnosis.

The other parasites listed typically do not cause bloody diarrhea.
A 25-year-old student is experiencing recurrent fevers and jaundice after returning from a trip to Sub-Saharan Africa. A diagnosis of *Plasmodium ovale* malaria is made. In addition to standard treatment, primaquine is added. Which stage of the infecting organism’s life-cycle is targeted by this drug?

- a. Gametocyte
- b. Hypnozoite
- c. Merozoite
- d. Sporozoite
- e. Trophozoite

**correct answer:** B

Malaria is a widespread tropical infection, causing disease in 400 million people per year, and killing 1-3 million. 4 species commonly cause malaria, *Plasmodium falciparum*, *P. malariae*, *P. ovale* and *P. vivax*. Of these, *P. vivax* and *P. ovale* may form hypnozoites, which can remain dormant in the liver for up to 30 years. Primaquine is effective at killing these sleeping (*hypno* = sleep) forms.
A 12-year-old girl with sickle cell disease has pain in her right arm. An x-ray of the right upper extremity shows bony lesions consistent with osteomyelitis. Which of the following is the most likely causal organism?

- a. Clostridium septicum
- b. Enterococcus faecalis
- c. Listeria monocytogenes
- d. Proteus mirabilis
- e. Salmonella enteritidis

**Correct answer:** F

You need to know the likely organisms that cause osteomyelitis, and determine which of these are associated with sickle cell disease.

In general, Staph aureus and Salmonella enteritidis are the two most common causes of osteomyelitis in general. The vaso-occlusive bone disease related to sickle cell leaves the bone prone to infection. Here, the best answer choice is (F) Salmonella enteritidis.

(A) Clostridium family bugs cause Gas Gangrene, myonecrosis and septicemia, NOT osteomyelitis.

(B) Enterococcus faecalis is a Gram-Positive bacterium that normally inhabits the human gastrointestinal tract. It can exhibit antibiotic resistance and cause endocarditis and bladder or prostate infections.

(C) Listeria monocytogenes is a Gram positive, motile (with flagella) organism. It causes the disease Listerosis, which is characterized by septicemia, encephalitis and uterine/cervical infections.

(D) Proteus mirabilis is a Gram-negative, facultatively anaerobic bacterium. It shows swarming, motility, and urease activity. So, Proteus infection commonly results in struvite or calcium carbonate crystals in the urine, and to the formation of kidney stones.
A 24-year-old male with a history of Sickle Cell disease presents to your office complaining of pain in his arms and legs of 12 hours duration. Laboratory tests identify that this patient is experiencing an ‘aplastic crisis.’ PCR tests demonstrate infection by a single stranded small DNA virus. This virus may also cause which of the following syndromes:

- a. Burkitt’s Lymphoma
- b. Erythema Infectiosum
- c. Molluscum Contagiosum
- d. Smallpox
- e. Yellow Fever

**correct answer:** B

In patients with sickle cell disease, aplastic crises are often caused by Parvovirus B19, a Single Stranded negative strand DNA virus. The structure and composition of such viruses provide important clues to diagnoses. Parvovirus B19 may also cause Erythema Infectiosum (fifth disease), a common exanthem in children, which often presents with a red “slapped cheeks” rash on the face. The rash is a result of a Th-1 and IgM mediated immune complex reaction to the virus, which deposits to form the exanthem.

Burkitt’s Lymphoma (A) may be long-term consequence of infection with Epstein Barr Virus (EBV), a type of Double Stranded Herpesvirus.

Molluscum Contagiosum (C) and Smallpox (D) may both result from Poxviruses, which are Double Stranded, Large DNA viruses.

Yellow Fever (E) is caused by infection with single stranded RNA viruses of the Flavivirus family.
A 2-year old child has a fever. She is irritable and tugs on her ear. Physical exam confirms otitis media, and laboratory analysis of the exudate shows a Gram-negative bacteria. Which is the most likely cause of the infection?

- a. Haemophilis influenzae
- b. Klebsiella pneumoniae
- c. Pseudomonas aeruginosa
- d. Streptococcus pneumoniae
- e. Streptococcus pyogenes

**Correct answer:** A

Pseudomonas aeruginosa causes malignant otitis externa but not otitis media; Streptococcus pyogenes and Streptococcus pneumoniae are Gram-positive; Klebsiella pneumoniae generally does not cause otitis media.
A patient being treated with clindamycin for aspiration pneumonia develops diarrhea. The stool contains a toxin that kills cultured epithelial cells. Stool culture grows an anaerobic gram-positive rod. The same organism is cultured from his bedpan. Which of the following is most likely to sterilize the bedpan?

a. Boiling for 45 minutes
b. Exposure to benzalkonium chloride for 1 hour
c. Exposure to ethyl alcohol for 1 hour
d. Exposure to saturated steam (121°C) for 15 minutes
e. Heating in an oven at 150°C for 30 minutes

**Correct answer:** D

The bacteria in question is Clostridium difficile (the most common cause of antibiotic-associated diarrhea). C. difficile is an anaerobic gram positive rod that forms spores. These spores are only destroyed by autoclaving (exposure to saturated steam for 15 minutes, option D), because the spore shell is resistant to other forms of sterilization.

As a side note, using alcohol based cleaners (like Purell) will not sterilize your hands after you see a patient with C. difficile infection. Therefore, it is important to physically wash your hands with soap and water, in order to mechanically wash the spores off your hands.
**Question 237**

**Clinical science: Multisystem/General**

Gram-positive cocci in clusters are seen in purulent fluid drained from a skin abscess. Rapid identification of these organisms will be facilitated by evaluation of the clumping of latex beads coated with which of the following?

a. IgG and fibrinogen
b. Interleukin-1 (IL-1) and factor VIII (antihemophilic factor)
c. Properdin and platelet factor 3
d. Prothrombin and C3b
e. Transferrin and plasminogen

**Correct answer:** A

In the question stem, the gram positive cocci in clusters are most likely Staph aureus and therefore, the answer choices must pertain to specific attributes of S. aureus. S. aureus expresses protein A which will bind to IgG and fibrinogen receptors (S aureus clumping factor) which will bind fibrinogen (option A)

IL-1 and factor VIII (B) are produced by the body and therefore will not identify S aureus.

Properdin and platelet factor 3 (C) are related to complement and platelet aggregation.

Prothrombin and C3b (D) are related to thrombin (coagulation cascade leading to clot formation) and complement factor part of immune response.

Transferrin is a iron carrier in the blood and plasminogen is precursor for plasmin which is an enzyme involved in fibrinolysis (clot busting).
After infection with measles virus, a 6-year-old boy produces antibodies to all eight viral proteins. The next year he is again exposed to measles virus. Antibodies to which of the following viral proteins are most likely to be protective?

a. Hemagglutinin  
b. Matrix  
c. Nonstructural  
d. Nucleocapsid  
e. Polymerase

**Correct answer:** A

Measles is an enveloped, single-stranded, negative sense RNA virus with a helical nucleocapsid. The hemagglutinin (HA) (choice A) viral protein is an envelope protein that binds sialic acid on cells allowing the virus to be endocytosed. Antibodies targeting viral envelope proteins have the best chance of binding their targets and providing the best immunity.

Matrix (choice B), nonstructural (choice C), nucleocapsid (choice D), and polymerase (choice E) proteins are shielded from their respective antibodies by the viral envelop and continue to be hidden once the virus is endocytosed into the host cell. Antibodies to the aforementioned proteins will not provide as much immunity from disease as antibodies to envelop proteins.
Hepatitis A virus (HAV) is resistant to solvents such as chloroform and 20% diethyl ether, while HIV-1 is sensitive to such solvents. This resistance is best explained by absence of which of the following in HAV?

a. Icosahedral symmetry  
b. Reverse transcriptase  
c. Single-stranded RNA  
d. Spherical structure  
e. Viral envelope

**Correct answer:** E

Chloroform and diethyl ether are organic solvents that break down lipid membranes, such as viral envelope (choice E). Enveloped viruses, like HIV-1, acquire their envelopes from the plasma membrane as they exit the cell they infected. This membrane is largely made up of lipids. Naked viruses, like HAV, lack this lipid viral envelope, and are therefore resistant to chloroform and diethyl ether. An important way of classifying viruses is whether or not they have an envelope.

The other answer choices do not affect whether the virus is sensitive to organic solvents.
Escherichia coli strains X and Y are both resistant to ampicillin. Ampicillin resistance is stable in strain X when it is grown for multiple generations in the absence of the antibiotic. However, strain Y loses ampicillin resistance when it is grown in media without the antibiotic. Which of the following best explains the acquisition of ampicillin susceptibility in strain Y?

a. Downregulation of the resistance gene  
b. Insertion of a transposon into the resistance gene  
c. Loss of a plasmid carrying the resistance gene  
d. Point mutations in the resistance gene  
e. Recombination with a defective copy of the resistance gene  

**correct answer:** C

Antimicrobial resistance in Escherichia coli is primarily conferred through transfer of a plasmid (choice C). This is true in the case of strains of Escherichia coli that carry a resistance to ampicillin. Without the need to produce the beta-lactamases needed to be resistant to ampicillin, it will not preferentially transfer the plasmid and it may be lost in subsequent generations.

The other four answer choices discuss changes to the actual gene which are not ways in which E. coli develops resistance.
Three weeks after a renal transplant, a patient develops fever and leukopenia, followed by prostration and severe pulmonary and hepatic dysfunction. Which of the following is the most likely viral cause?

a. Adenovirus type 12
b. Coxsackievirus
c. Cytomegalovirus
d. Influenza virus
e. Parvovirus B19

Correct answer: C

Cytomegalovirus (CMV) infection in immunocompetent hosts is usually subclinical, or at most presents as a mild form of mononucleosis similar to Epstein-Barr virus with mild fever, lymphadenopathy, splenomegaly and atypical circulating lymphocytes for several weeks. However, in immunocompromised hosts, such as the post-transplant patient above, CMV infection or reactivation of a latent CMV infection can manifest as a multisystem disease within 20-60 days. In addition to the aforementioned features of mononucleosis, CMV in immunocompromised hosts also manifests as pneumonia, hepatitis, gastritis and colitis. In late stage AIDS patients, CMV particularly causes a severe form of retinitis.

Adenovirus also causes a worsened form of its normal disease in immunocompromised hosts, but usually presents with severe gastroenteritis (vomiting and diarrhea) and pneumonia/bronchitis/ARD. Coxsackie virus usually presents in childhood as hand-foot-and-mouth disease, causing painful, vesicular, blisters in the oral mucosa, tonsillar pillars and palms and soles. Though influenza may cause a severely fatal pneumonia in immunocompromised hosts, it does not cause usually cause liver abnormalities. Parvovirus usually causes a chronic infection of the bone marrow and should be suspected in post-transplant patients with erythropoietin-resistant anemias.
Three weeks after traveling to California to study desert flowers, a 32-year-old man develops a fever, chest pain, and sore muscles. Two days later, red tender nodules appear on the shins, and the right ankle is painful and tender. An x-ray of the chest shows a left pleural effusion. Which of the following is the most likely diagnosis?

a. Blastomycosis  
b. Coccidioidomycosis  
c. Histoplasmosis  
d. Mycobacterium marinum infection  
e. Mycoplasma pneumoniae infection

**correct answer: B**

When the USMLE question involves a traveler returning to a trip from a specific locale, it is usually a clue to the diagnosis. In this case, coccidiomycosis is a well-known fungal disease endemic to California, Arizona, New Mexico and Nevada, giving it the moniker "Valley fever." It usually presents several weeks after exposure as a flu-like illness with fever, myalgias and erythema nodosum over the shins. Patients who fail to clear the infection may progress to chronic pulmonary infection, pulmonary effusions or meningitis.

Blastomycosis and Histoplasmosis are also systemic fungal infections, but the travel history usually involves a recent visit to the Mississippi/Ohio River valley. M. pneumoniae causes a pneumonia syndrome, and M. marinum causes systemic skin nodules, but both are usually opportunistic infections of the immunocompromised.
A 45-year-old woman develops a high spiking fever and shaking chills 2 weeks after returning from central Africa. Which of the following tests on the patient’s blood will establish the most likely diagnosis?

- a. Acid-fast stain
- b. Giemsa stain
- c. Gram stain
- d. India ink wet mount
- e. KOH stain

**Correct answer:** B

A high spiking fever and shaking chills after a trip to central Africa is a classical story for malaria of the Plasmodium genus. The conventional method for diagnosing malaria is a (B) Giemsa-stained blood smear.

(A) Acid-fast stains are classically used for diagnosis of tuberculosis, while (D) India ink wet mount is known for its use with Cryptococcus neoformans. (E) KOH stain is used for fungal elements in thick mucoid material and in specimens containing keratinous material, such as skin scales, nails, or hair.
A 34-year-old man develops a flu-like illness that is accompanied by a red, flat, round rash on his leg that progressively expanded outward over the course of a week. As the rash expanded, the center of the rash slowly turned clear. What was the most likely event that introduced the pathogen for this patient’s presentation?

a. Airborne droplet inhalation
b. Fecal-oral ingestion
c. Needle-stick
d. Tick bite
e. Unprotected sexual contact

**Correct answer:** D

The rash described is erythema chronicum migrans (ECM), the primary lesion of Lyme disease, which is caused by Borrelia burgdorferi. A tick bite (choice D) from the Ixodes tick transmits the spirochete from the white-footed mouse reservoir, and the illness starts about 10 days after the bite with a single ECM rash, a flu-like illness, and regional lymphadenopathy.

Lyme disease is not transmitted via airborne droplets, fecal-oral, needle-sticks, or unprotected sexual contact.
A 66-year-old man presents with chronic leg swelling. He is a recent immigrant from Fiji. He reports episodes of fever and headache. Physical exam reveals thick, scaly skin on both legs with swelling extending to the genitals. Swollen inguinal lymph nodes are palpable. What was the likely vector of transmission for his infection?

a. Flea
b. Body Louse
c. Mite
d. Mosquito
e. Tick

**Correct answer:** D

This man exhibits the signs and symptoms of elephantiasis, a condition caused by the nematodes Wuchereria bancrofti and Brugia malayi. They are both spread by the bite of an infected mosquito (choice D). The lymphatics are infected, resulting in chronic leg and genital swelling. Wuchereria infection is endemic to the Pacific Islands, much of Africa, whereas Brugia is found in the Malay Peninsula and much of Southeast Asia. Importantly, mosquitoes are vectors for many other bacteria species, which you should be familiar with.

Fleas (A) may serve as vectors for the transmission of Yersinia pestis (bubonic plague) between rodents and humans. Murine typhus (endemic typhus) fever, and in some cases Hymenolepiasis (tapeworm) can also be transmitted by fleas.

Body Lice (B) serve as vectors for Rickettsia prowazekii, causing Epidemic typhus, Bartonella quintana (trench fever causing endocarditis commonly among the homeless), and Borrelia recurrentis (a recurrent fever found in east Africa).

Mites (C) bites may cause otitis or dermatitis and can act as vectors for Serratia marcescens or Staphylococcus intermedius/Staphylococcus chromogens.

The tick (E) can act as a vector for Borrelia Burgdorferi or Coxiella-like species. Most famously, tick bites carrying Borrelia are the cause of Lyme Disease.
A 61 year-old diabetic male presents with facial pain on the right side, progressively worsening over the past 5 days. The maxillary sinus is biopsied and reveals the image above.

This organism is associated with which of the following?

- a. Diabetic ketoacidosis
- b. Misuse of common antibiotics
- c. HLA B27 haplotype
- d. Exposure to bird or bat droppings
- e. Live vegetation with budding yeasts

**Correct answer:** A

The correct answer is A. The patient may have diabetic ketoacidosis, which is a risk factor associated with mucormycosis. Mucor is identified in the image by broad, nonseptate hyphae with 90 degree branching. This mold may invade sinus cavities and cause sinusitis as in this patient.

Choice B is incorrect; overuse or misuse of antibiotics is not a risk factor for mucormycosis.

Choice C is incorrect; HLA B27 haplotype is associated with seronegative spondyloarthropathies such as ankylosing spondylitis, Reiter's syndrome, sacroiliitis, and psoriatic arthritis.

Choice D is incorrect; exposure to bird or bat droppings is classically associated with histoplasmosis.

Choice E is incorrect; exposure to live vegetation (with budding yeasts) is the cause of cases of sporotrichosis, which is a dimorphic fungus that causes nodules along draining lymphatics, most commonly along the wrist and forearm. This is also known as "rosegardener's disease."
A 23-year-old man presents to your office with a five day history of unremitting fever and right wrist pain. He denies a history of illicit drug use. Vitals are as follows: HR 118, BP 135/90, RR 20, T 101.9F. The patient is noticeably anxious. On physical exam, his right wrist and left elbow are both painful to palpation. No needle marks are visible. Small papular, non-pruritic lesions are noted on his back. Which of the following is the likely event that introduced the pathogen for this patient’s presentation?

a. Airborne droplets
b. Fecal-oral transmission
c. Needle stick injury
d. Operative injury
e. Unprotected sexual contact

Correct answer: E

This patient is presenting with likely gonococcal arthritis given his young age, polyarticular symptoms, fever and rash. It is likewise probable that he was infected during unprotected sexual contact with an individual exhibiting genital gonorrhea. Our patient was unable to contain the organism locally, either due to a failure in his own immunity or the increased virulence of the gonococcal strain, and he presented with the arthritic form.

Airborne droplets can create an osteomyelitis-type picture in the case of TB-induced Pott's disease of the vertebrae, particularly in indigent populations in developing countries. However our patient fits none of the risk factors or symptoms for TB. Fecal-oral transmission may create osteomyelitis in the case of Salmonella, but this is particular in patients who are already afflicted with sickle cell disease. Needle sticks and operative injury may introduce myriad organisms that can cause septic arthritis (i.e. Staph aureus, Pseudomonas aeruginosa), but our patient has no history of this type of introduction.
A 22-year-old college student that is confused and lethargic is brought to the emergency room. His roommates say he was complaining of a severe headache and neck stiffness last night. Vital signs are remarkable for a temperature of 104.3, blood pressure of 80/60 and heart rate of 120. Physical exam reveals a purpuric rash on the patient’s chest. Lumbar puncture reveals a high neutrophil count.

What treatment should be given to his roommates as prophylaxis?

a. Acyclovir
b. Ceftriaxone
c. Doxycycline
d. Rifampin
e. Vancomycin

**correct answer:** D

The patient described has acute bacterial meningitis, the cause of which is most likely N. meningitidis (given the patient’s age, clinical history, and the characteristic purpuric rash of DIC.) Rifampin is indicated for prophylaxis of close contacts of patients with meningococcal meningitis.

Ceftriaxone is indicated for the treatment of meningococcal meningitis, but not the prophylaxis of contacts.

Vancomycin is not indicated in this situation as it only covers gram positive organisms.

Acyclovir is also not indicated in this situation as the lumbar puncture results suggest this patient’s meningitis is bacterial and not due to a virus.

Doxycycline is not indicated for the prophylaxis of meningococcal meningitis.
A 26-year-old HIV-positive woman is brought into the emergency room after she is found lying unresponsive in the street. She has no recollection of what had occurred. She reports sensing a smell "like burning hair" and then blacking out. A medical history reveals that she has had a fever and a headache for the past week. An MRI of the brain shows multiple ring-enhancing lesions. Which of the following is the most likely cause of her symptoms?

a. Cryptococcus neoformans
b. Cryptosporidium
c. Cytomegalovirus
d. Pneumocystis carinii
e. Toxoplasma gondii

**Correct answer:** E

This patient has likely suffered a seizure due to an infection of Toxoplasma in her brain. The ring-enhancing necrotic lesions in the brain are highly characteristic of toxoplasmosis. HIV-infected and other immunocompromised patients are vulnerable to brain infections by Toxoplasma, which is spread by cysts in meat or cat feces.

The other microorganisms listed are also opportunistic infections which affect HIV-positive patients, but they do not cause ring-enhancing brain lesions.

A non-infectious cause of ring-enhancing brain lesions in AIDS patients is B cell lymphoma of the brain.
A 35 year old male with human immunodeficiency virus comes to the ER presenting with one week of fever, malaise, and headache. He also reports having some neck stiffness over the past two days. On physical exam, he has a temperature of 102.4 F with stable blood pressure and heart rate. He has no focal neurological deficits, but is very uncomfortable when a pen light is shined in his eye to examine his pupils. You suspect cryptococcal meningitis and perform a spinal tap to obtain CSF. Which preparation would most likely lead to the correct identification of the organism responsible for this patient’s meningitis?

- a. Congo red
- b. India ink
- c. Periodic acid-Schiff
- d. Silver stain
- e. Ziehl-Neelsen

**Correct answer:** B

India ink (choice B) will correctly stain for cryptococcus meningitis.

Congo red (choice A) will reveal amyloid that is characterized by apple-green birefringence in polarized light.

Periodic acid-Schiff (choice C) stains glycogen and mucopolysaccharides. It is commonly used to diagnose Whipple’s disease.

Silver stain (choice D) is used to stain fungi such as pneumocystis carinii. It is also used to stain for legionella.

Ziehl-Neelsen (choice E) is used to stain acid-fast bacteria.
A 25-year-old sexually active female with no significant medical history presents to the clinic complaining of urinary frequency and dysuria for the past 3 days. Physical exam is unremarkable. Urinalysis reveals presence of leukocyte esterase and nitrites.

What is the most likely causative organism?
- a. Chlamydia trachomatis
- b. Escherichia coli
- c. Haemophilus ducreyi
- d. Pseudomonas aeruginosa
- e. Serratia marcescens

**correct answer: B**

This patient's history and laboratory testing are consistent with urinary tract infection (UTI). Leukocyte esterase indicates presence of white blood cells responding to a bacterial infection; Gram negative organisms cause production of nitrites. Escherichia coli (choice B) fits these characteristics and is the most common cause of UTI in ambulatory women.

Chlamydia trachomatis (choice A) can cause urethritis, which would present with dysuria and urethral discharge. Absence of discharge and presence of urinary frequency suggest cystitis rather than urethritis. Additionally, chlamydia is not associated with nitrite production.

Haemophilus ducreyi (choice C) does not cause UTI. Symptoms include painful ulcers on the genitals and inguinal adenopathy.

Pseudomonas aeruginosa (choice D) UTI is usually nosocomial, and therefore unlikely in this ambulatory patient. Additionally, incidence of E. coli UTI is far greater.

Serratia marcescens (choice E) UTI is usually nosocomial, and therefore unlikely in this ambulatory patient. Additionally, incidence of E. coli UTI is far greater.
A 19-year-old college student presents to the sexual health clinic with a painless open lesion on his penis. An antibiotic is prescribed and the patient’s symptoms soon disappear. The most likely antibiotic functions by interfering with which of the following cellular components?

a. 30S ribosome
b. 50S ribosome
c. Cell wall peptidoglycans
d. DNA gyrase
e. Dihydrofolate reductase enzyme

**correct answer:** C

This student has the classic presentation of "chancre" from primary syphilis. The treatment of choice is still Penicillin G in non-allergic patients. Penicillins work by blocking formation of the cell wall that is made up of cross-linked peptidoglycans. Trimethoprim and pyrimethamine are folate inhibitors that block DHFR. Folate is needed for DNA replication of bacteria. Fluoroquinolones block DNA gyrase, needed to transcribe bacterial proteins. Macrolides and clindamycin block the 50S ribosome needed to elongate bacterial proteins. Aminoglycosides and tetracyclines block the 30S ribosome.
A 1-day-old newborn is evaluated for possible sepsis. Blood cultures grow gram-positive cocci in pairs and chains that agglutinate with group B antiserum. The most likely epidemiologic risk factor for this infection involves bacterial colonization of which of the following?

- a. Mother’s vagina
- b. Newborn's gastrointestinal tract
- c. Newborn's nasopharynx
- d. Placenta
- e. Umbilical cord remnant

**Correct answer:** A

The description of the bacteria is significant because it identifies the organism as group B streptococcus. Group B strep is a very common cause of newborn meningitis and sepsis, and it is most often found colonizing the mother’s vagina (option A).

The newborn's GI tract (B) is not completely sterile, but group B strep is not part of the flora.

Colonization of the newborn’s nasopharynx (C) is incorrect because the infection is not transmitted by the respiratory route, nor is it transplacental infection (D) (for transplacenta think TORCH), nor would it colonize the umbilical cord remnant (E) (an infection here would most likely be from an anaerobic bacteria).
A newborn develops meningitis. *Streptococcus* is isolated from the mother’s vagina. The organism agglutinates with antiserum directed against type B surface carbohydrate. The virulence of this organism is related to a bacterial constituent that interferes with which of the following host phagocyte functions?

- a. Aggregation
- b. Chemotaxis
- c. Ingestion
- d. Intracellular killing
- e. Pseudopod formation

**Correct answer:** C

Group B Streptococci, or more specifically *S. agalactiae*, occasionally colonize the female reproductive tract and are common causes of neonatal meningitis. The best characterized virulence factors of Group B Streptococci are the capsular polysaccharides, which confer serotype specificity. The capsule provides virulence by inhibiting the deposition of complement components on the surface of the organism and therefore inhibiting ingestion (choice C) by cells of the immune system.

The other answer choices are examples of virulence factors of other organisms, but not Group B Streptococci.
A 30-year-old man comes to the clinic because of a painful ulcer on his penis for the past week. He has had multiple sexual partners, including commercial sex workers. Physical examination shows lymphadenopathy in the inguinal region and a 1-cm tender ulcer with no induration located on the frenulum. A culture of the ulcer grows colonies on supplemented chocolate agar. A Gram stain of the colonies shows gram-negative coccobacilli. Which of the following is the most likely causal organism?

a. Haemophilus ducreyi
b. Herpes simplex virus
c. Neisseria gonorrhoeae
d. Treponema pallidum
e. Trichomonas vaginalis

**correct answer: A**

Haemophilus ducreyi (choice A) is a Gram-negative coccobacillus that is typically grown on chocolate agar. Like the other answer choices, it is a sexually transmitted disease, but it is characterized by painful lesions in the genitalia.

Herpes simplex virus (choice B), like other viruses, cannot be Gram stained. The test of choice for identifying a herpes virus is a Tzanck test.

Neisseria gonorrhoeae (choice C) is a Gram-negative diplococci.
Treponema pallidum (choice D) is the bacteria that causes syphilis and is a spirochete.

Trichomonas vaginalis (choice E) parasitic flagellated protozoan.
A 29 year old sexually active woman comes into your office complaining of a “fishy smelling” discharge coming from her vagina for the past few days. She denies any dysuria or inflammation. She has been diagnosed and treated for gonorrhea in the past. On exam, you identify a thin, grayish-white discharge that coats the vaginal walls. You take a culture and send it to the lab. It is reported that there are clue cells on saline wet mount. What organism is most likely responsible for this woman’s symptoms?

a. Chlamydia trachomatis
b. Gardnerella vaginalis
c. Neisseria gonorrhoeae
d. Staphylococcus aureus
e. Streptococcus agalactiae

Correct answer: B

G. vaginalis (choice B) is a pleomorphic rod that causes bacterial vaginosis. It is typically found in sexually active women and is commonly asymptomatic. The most common presenting symptom is a homogenous, thin, grayish-white discharge from the vagina. The diagnosis is made by the identification of clue cells on saline wet mount. Clue cells are vaginal epithelial cells covered with bacteria.

C. trachomatis (choice A) and N. gonorrhoeae (choice C) are common sexually transmitted infections, but these would most likely present with a painful discharge and there would most likely be no clue cells.

S. aureus (choice D) is part of the normal skin flora. It can cause inflammatory processes such as skin infections or abscesses, as well as toxin-mediated disease. Toxic shock syndrome should be suspected in women who wear tampons when menstruating.

S. agalactiae (choice E), also known as group B streptococci, frequently colonizes the human genital tract. It can be vertically transmitted from mother to child, causing neonatal sepsis and meningitis.
A 23 year sexually active woman presents with a right upper quadrant pain of one day duration. Her pelvic exam is remarkable for purulent vaginal discharge and cervical motion tenderness. Speculum exam reveals a greening material coming out of a reddened cervix. What is the explanation of her RUQ pain?

a. Distention of the liver capsule  
b. Choledocholithiasis  
c. Cholelithiasis  
d. Pyelonephritis  
e. Bacterial vaginosis

**Correct answer:** A

This patient has Fitz-Hugh-Curtis syndrome, characterized by RUQ pain secondary to distention of the Glisson’s capsule of the liver. This condition is a rare sequelae of pelvic inflammatory disease (PID), in which chronic gonococcal or chlamydial inflammation causes adhesions and scar tissue to form on the liver capsule, which distends it. It occurs almost exclusively in women. Any purulent discharge in a sexually active patient should raise suspicion about gonorrhea. Although PID may be asymptomatic, it can present as one or more of the following symptoms: fever, lower abdominal pain, cervical motion tenderness, discharge, and painful intercourse.

B is incorrect. Choledocholithiasis refers to stones in the common bile duct. It may cause RUQ pain, but does not explain the vaginal discharge and cervical motion tenderness.

C is incorrect. Cholelithiasis refers to stones in the gallbladder, which may cause RUQ pain, but not cervical motion tenderness.

D is incorrect. Pyelonephritis is infection of the kidney, which may cause RUQ pain. However patients usually have fever. The full picture of the case is not accounted by pyelonephritis.

E is incorrect. Bacterial vaginosis is diagnosed when at least 3 out of 4 criteria are satisfied (Amsel criteria)

1. Thin, gray white vaginal discharge  
2. Vaginal pH > 4.5  
3. Positive Whiff test (release of fishy odor on adding alkali—10% potassium hydroxide (KOH) solution)  
4. Clue cells (epithelial cells covered by bacteria)
A 5 year old boy is brought into the emergency department late in the evening by his mother who reports that her child is having difficulty breathing. The mother believes that the child swallowed a small toy he was playing with before bed and insists the boy was completely normal before being put to bed earlier that night. Physical exam reveals a child in respiratory distress, sitting upright and forward with audible inspiratory stridor. Inspection of the oropharynx is clear. In addition, the child is febrile to 102.5ºF and is drooling from the mouth. What is the most likely etiology?

- a. Foreign body obstructing upper airway
- b. H. influenza acute epiglottitis
- c. Compromised airway from child abuse
- d. RSV upper respiratory infection
- e. Streptococcus upper respiratory infection

correct answer: B

H. influenza (choice B) can cause multiple conditions including acute epiglottitis, meningitis, otitis media, and pneumonia. In the setting of a fever, inspiratory stridor, and drooling from the mouth, the diagnosis of acute inflammation of the epiglottis causing obstruction of the airway needs to be recognized. Lateral X-rays of the neck can be used to confirm the clinical diagnosis.

Foreign body obstructing the airway (choice A) would not explain the high fever that the child is experiencing.

Compromised airway from child abuse (choice C) is not likely in the setting of a fever. Additionally, bruising patterns on the child’s neck suggesting choking were not described on physical exam.

RSV (choice D) is not a known cause of epiglottitis; however, it can cause respiratory tract infections (bronchiolitis, pneumonia) in children and infants.

Streptococcus (choice E) does not commonly cause epiglottitis but is more commonly the cause of exudative pharyngitis which was not described on physical exam.
A 45 year old man with HIV (CD4 count 180) presents with difficulty breathing, weight loss, and low grade fevers. A chest x-ray shows bilateral diffuse interstitial infiltrates. Bronchoalveolar lavage demonstrates PCP pneumonia. The gentleman notes that he is allergic to sulfa, how should he best be treated?

- a. TMP-SMX
- b. Pentamidine
- c. Doxycycline
- d. Amphotericin B
- e. Itraconazole

**Correct answer:** B

In patients with PCP pneumonia who are allergic to sulfa TMP-SMX cannot be used safely. The best alternative treatment in these patients is Pentamidine. While it is not the first line therapy in patients who are not sulfa-allergic, it can successfully be used in patients with sulfa-allergy. Remember that HIV patients with CD4 counts less than 200 need TMP-SMX for prophylaxis against both PCP Pneumonia and toxoplasmosis. The other drugs have no effect on PCP pneumonia. Know how to treat the common opportunistic infections in AIDS: PCP, toxo, TB, and MAC.
A 35-year-old recent immigrant from India presents to the clinic with new well-defined hypopigmented macules on his back and left arm. The patient also reports decreased sensation of affected areas and on the fingers of the left hand. What is the most likely result of skin testing for bacterial pathogens of this patient’s hypopigmented lesions?

a. Abundant acid-fast bacilli  
b. Abundant gram positive cocci in clusters  
c. Few to no acid-fast bacilli  
d. Positive India Ink stain  
e. Positive growth on MacConkey’s agar

**Correct answer:** C

While uncommon in the West, India, Nepal and Brazil have the highest prevalence of Leprosy, and this case presents the typical findings of tuberculoid leprosy: anesthetic hypopigmented macules with distal sensory nerve damage. If skin lesions are tested for the causative organism, Mycobacterium leprae, the lesions will typically yield few to no acid-fast bacilli (C). In contrast, lesions of lepromatous leprosy are characterized by extensive tissue destruction, leonine facies, and diffuse nerve involvement, and will reveal abundant acid-fast bacilli (A). The other answers are incorrect because mycobacteria do not stain through gram staining (B), will not stain positive with the special fungal stain India Ink (D), and are fastidious and will not grow on MacConkey’s agar (E). Classically staphylococci will appear as gram positive cocci in clusters, gram negative enterics such as E. coli will grow on MacConkey’s agar and Cryptococcus stains positive with India Ink.
An asymptomatic 66 year-old man has a routine complete blood count which reveals a leukocytosis of 50,000 white blood cells per microliter. On fluorescent in situ hybridization assay, he is found to have the Philadelphia chromosome in peripheral blood mononuclear cells, and he is diagnosed with chronic myelogenous leukemia. Which of the following is the mechanism of action for the most effective initial drug therapy for this disease?

a. An antibody that binds to and inhibits Her2/neu on the cell surface.
b. An antibody that binds to and inhibits CD20 molecules on the surface of B cells.
c. A small molecule that binds to and inhibits the kinase domain of the BCR-Abl protein.
d. A small molecule that binds to and inhibits the kinase domain of the epithelial growth factor receptor (EGFR).
e. A molecule that binds to tubulin dimers and hence inhibits the assembly of microtubules during mitosis.

correct answer: C

The answer is C. The patient with Philadelphia chromosome-positive CML should first be treated with imatinib (Gleevec), which is a small molecule tyrosine kinase inhibitor that binds to the BCR-Abl protein. It inhibits the protein by binding its kinase domain and decreasing the protein's affinity for ATP. This protein is the result of a chromosomal translocation occurring in CML which fuses chromosomes 9 and 22 and leads to production of an abnormal oncogenic protein, BCR-Abl. This drug has also proven to be effective in some gastrointestinal stromal tumors (GIST) that express mutated c-Kit protein.

Choice A is incorrect; this choice describes trastuzumab (Herceptin) which is commonly used in breast cancers that overexpress Her2/neu (also known as ErbB2).

Choice B is incorrect; this choice describes rituximab which is sometimes used in B cell lymphoma and other diseases.

Choice D is incorrect; this description is true of both gefitinib (Iressa) and erlotinib (Tarceva) and these drugs are sometimes used in non-small-cell lung cancers.

Choice E is incorrect; this choice describes vincristine, a vinca alkaloid which is usually part of multi-drug regimen for certain cancers, usually non-Hodgkin's lymphoma.
A 6 month-old male is thought to have sickle cell anemia, and as his pediatrician you want to sequence the gene that may be mutated, causing this disease. You decide to sequence the gene for the beta subunit of hemoglobin. In order to do this you must perform polymerase chain reaction (PCR). You need to know which of the following in order to amplify (PCR) the area of interest of his DNA?

a. The predicted change in nucleotide sequence
b. The nucleotide sequence in the areas upstream and downstream to the nucleotides of interest
c. The predicted change in amino acid sequence of the mutated protein
d. The change in number of hydrogen bonds within the nucleotide sequence of interest
e. The predicted change in size of the mutated protein

**Correct answer:** B

**Answer:** B. The answer is B. Polymerase chain reaction is a method with which to amplify an area of interest of DNA. Normally, specifically designed primers bind upstream and downstream to the area of interest. The sequence in between these primers is amplified and then sequenced to look for specific nucleotide changes that will then predict a possible change in amino acid sequence and protein structure and function. In sickle cell anemia, the beta subunit of hemoglobin is mutated at the 6th codon, and glutamate (wild type) is replaced by valine in the protein. This missense mutation changes protein function and causes sickle cell anemia which is inherited in an autosomal recessive manner.

Choice A is incorrect; the predicted change is unnecessary for performing PCR and sequencing the area of interest. The only known sequences must be where the primers bind to DNA.

Choice C is incorrect; polymerase chain reaction deals with nucleic acids, DNA in this case. Protein sequence is then predicted by changes in the DNA nucleotide sequence.

Choice D is incorrect; predicted changes in numbers of hydrogen bonds may influence the parameters of the PCR assay itself but have no influence on nucleotide or amino acid sequence.

Choice E is incorrect; PCR is done on DNA and thus the predicted size of the protein does not influence the assay itself.
A scientist wishes to insert her gene of interest into a yeast vector. She decides to use a restriction endonuclease that cleaves the vector at a palindromic recognition site leaving sticky ends. Which of the following is a possible recognition site for the restriction enzyme?

a. 5'-AAGCTA, 3'-TTCGAT
d. 5'-GGATCG, 3'-CCTA

correct answer: C

d. 5'-GCATTC, 3'-CGTAAG
e. 5'-CAATTG, 3'-GTTAAC

c. 5'-CAATTG, 3'-GTTAAC

e. 5'-TCAGGA, 3'-AGTCCT

Don't worry -- you don't need to memorize the recognition sequences! You simply need to know that restriction enzymes that splice at recognition sites are almost always palindromic. There are some restriction enzymes that cut outside the recognition site; these do not necessarily have palindromic recognition sites. Here, 5'-CAATTG & 3'-GTTAAC is the only answer choice with a pallindromic sequence.
The diagram shows the equilibrium between active and inactive forms of a receptor (R), its interaction with a drug (V), and the coupling of the active form of the receptor to a signal transduction pathway. Drug W acts at site Y of the signal transduction pathway to diminish the physiologic effect of drug V. Which of the following terms accurately describes the effect of drug W on the dose-response curve for drug V?

- a. Competitive antagonism
- b. Full agonism
- c. Noncompetitive antagonism
- d. Partial agonism
- e. Reverse agonism

**Correct answer:** C

A noncompetitive antagonist (C) binds at a different site than the receptor, or irreversibly binds the receptor site. In either case, the system is not activated. Thus, response is lowered at a given dose of agonist, and this effect is not overcome with increased agonist dose. (The dose-response curve shifts downward.) Drug W meets this description.

A competitive antagonist (A) reversibly binds to a receptor without activating the system, lowering the response at a given agonist dose. However, a response of maximum efficacy may be achieved with increased agonist dose. (The dose-response curve shifts to the right.) Drug W is not a competitive antagonist because it acts at a site other than the receptor for drug V. Further, it is not stated that the effect of drug W is overcome with increasing doses of drug V.

A full agonist (B) will produce a response of maximum efficacy if present in sufficient quantity. Drug W cannot be a full agonist, because it diminishes the physiologic effect of drug V.

When administered alone, a partial agonist (D) binds to the receptor site and elicits a less-than-100% response regardless of dose. When administered with a full agonist, it acts like an antagonist because it competes for receptor binding sites. (The dose-response curve shifts downward, and may also shift left or right.) Drug W cannot be a partial agonist because it acts at a site other than the receptor for drug V.

A reverse agonist (or inverse agonist) (E) binds to the same receptor site as an agonist, but elicits the opposite effect. This choice is incorrect because drug W does not bind at the same site as drug V, and only a diminished (rather than opposite) effect is observed.
Several contiguous cells are labeled with a fluorescent dye that cannot cross cell membranes. One cell is experimentally bleached with light that destroys the dye, but the cell soon recovers dye fluorescence. This recovery is best explained by the presence of which of the following structures between the bleached cell and its fluorescent neighbors?

a. A basal lamina
b. Desmosomes (maculae adherentes)
c. Gap junctions
d. Glycosaminoglycans
e. Tight junctions (zonulae occludentes)

**Correct answer:** C

Gap junctions (choice C) allow transfer of chemical and electrical signals between cells. The key structural component of gap junctions is the connexon, a hollow cylinder made up of transmembrane proteins. Gap junctions allow cells to coordinate activities with each other, and are found in nerve cells, cardiac muscle, smooth muscle, and liver cells.

Basal lamina (choice A) is a layer of extracellular matrix on which the epithelium sits.

Desmosomes (choice B) are structures that bind to intermediate filaments between adjacent cells in order to hold them together.

Glycosaminoglycans (choice D) are the carbohydrate portions of proteoglycans, or mucopolysaccharides, that form the major structural components of the extracellular matrix.

Tight junctions (choice E) are barriers that form a belt-like structure encircling epithelial cells along their apical surface. They prevent movement of small molecules between the two cells.
A 12-year-old girl is admitted to the hospital because of marked shortness of breath, an erythematous rash, and painful, swollen hip and knee joints. She is agitated. A chest x-ray shows an enlarged heart and changes consistent with pulmonary edema. Intractable congestive heart failure develops, and she dies on the second hospital day. This child most likely had a recent history of which of the following?

- a. Cyanosis with chest pain
- b. Jaundice
- c. Meningitis
- d. Pharyngitis
- e. Skin infection

**Correct answer:** D

This girl has severe carditis, which resulting enlargement of the cardiac shadow and acute congestive heart failure. This is a severe consequence of rheumatic fever, which is suggested by the acute-onset polyarthritis and rash (erythema marginatum). Rheumatic fever generally occurs 1-5 weeks after an upper respiratory infection caused by beta-hemolytic streptococcus. It is postulated that the immune system's response to the strep results in attack also on native tissues with similar antigens ("molecular mimicry").

Averting the possibility of rheumatic fever after streptococcus infection is the basis of penicillin treatment and prophylaxis for all people with suspected infection.
A 86-year-old female patient is being evaluated for fatigue, altered mental status and ataxia at her physician's office. Physical exam is notable for loss of proprioception, and laboratory values are notable for Hb of 8.9 g/dl with a MCV of 103. A blood smear is notable for macrocytic erythrocytes and hypersegmented neutrophils. Which of the following will most likely be elevated in the blood?

a. Ferritin
b. HbS
c. LDH
d. Methionine
e. Methylmalonyl-CoA

**correct answer:** E

This patient is suffering from B12 deficiency which is manifesting as macrocytic anemia with mental status changes and ataxia due to lack of proprioception. B12 is a necessary cofactor in the conversion of methylmalonyl Co-A into Succinyl-CoA (entering the Kreb cycle) and therefore methylmalonyl CoA will build up.

Ferritin is increased in anemia of chronic disease, but there is no sign of a chronic disease in this patient and anemia of chronic disease is usually normocytic.

HbS is seen in sickle cell disease which would be rare in an 86 year old and a smear would most likely show sickle cells.

LDH is elevated in hemolytic anemia which is another type of normocytic anemia.

Methionine is the compound that is regenerated from homocysteine via B12 and THF (active folate). While homocysteine would be elevated, methionine would be decreased in B12 deficiency (and folate deficiency).
A researcher has discovered a rare and previously unknown autoimmune disease. CD8+ T cells isolated from patients are resistant to apoptosis, and proliferate at a much higher rate than normal. Upon further study, the researcher discovers that these cells contain a mutation in a cell surface receptor. The mutation causes the receptor to become inactive. However Western blotting shows that the mutant and wild-type receptors have the same molecular weight. What type of mutation do these patients carry?

a. Deletion mutation
b. Frameshift mutation
c. Missense single-nucleotide mutation
d. Nonsense single-nucleotide mutation
e. Splice site mutation

**Correct answer:** C

The only mutation which would result in a protein of the same molecular weight is a missense mutation which causes a single amino acid substitution. This may cause a structural change in the receptor, making it inactive or unable to bind its ligand.

Deletion, nonsense and frameshift mutations would all result in a shortened protein.

A splice site mutation causes an intron to be left in during mRNA processing, resulting a longer protein (or a shorter protein if the intron contains a stop codon).
A 13-year-old female presents with her mother requesting information about the mechanism by which human papillomavirus (HPV) increases the risk of cervical carcinoma. Which of the following statements is true regarding HPV-associated cervical carcinoma?

a. E6 activates E2F-mediated transcription
b. E7 inhibits E2F-mediated transcription
c. E6 activates the retinoblastoma (Rb) gene product
d. E7 inhibits the retinoblastoma (Rb) gene product
e. E6 activates the tumor suppressor p53

Correct answer: D

The carcinogenic effects of HPV types 16 and 18 are primarily associated with viral antigens E6 and E7. E6 promotes cervical carcinoma through inhibition of the tumor suppressor p53. E7 does so via inhibition of the retinoblastoma (Rb) gene product, which disinhibits E2F and promotes transcription and cell proliferation.

E6 does not influence E2F-mediated transcription (A); rather, E6 works by inhibiting p53.

E7 activates E2F-mediated transcription as described above; it does not inhibit it (B).

E6 does not activate the Rb gene product (C); rather, it inhibits p53.

E6 inhibits p53; activation of p53 (E) would decrease the risk of carcinoma, it would not promote carcinogenicity.
A 62-year-old man presents to his primary care physician complaining of a recent eruption of blisters all over his body. On exam, the patient has multiple tense bullae in his axillae, groin and in flexural areas of his arms bilaterally as seen in the accompanying image. His oral exam reveals pink mucosa with no lesions. The physician takes a punch biopsy at the site of one of the blisters, which demonstrates deposits of IgG and complement along the basement membrane of the epidermis. This patient’s symptoms likely represent an autoimmune reaction targeting which of the following structures?

a. Desmosome  
b. Hair Follicle  
c. Hemidesmosome  
d. Melanocyte  
e. Pacinian corpuscle

**Correct answer:** C

This patient has bullous pemphigoid, an autoimmune blistering disorder. In this disease, autoantibodies are formed against basement membrane hemidesmosome proteins. The hemidesmosome is a specialized structure that anchors cells to the extracellular matrix; in the skin, hemidesmosomes attach keratinocytes to the basement membrane. The destruction of hemidesmosomes in bullous pemphigoid results in the formation of subepidermal blisters or “tense bullae” as opposed to the flaccid bullae seen with pemphigus vulgaris. On punch biopsy, a characteristic deposition of IgG and complement can be seen along the basement membrane.

A. Proteins in the desmosome are targeted in the autoimmune blistering disorder known as pemphigus vulgaris. The desmosome is a specialized structure which anchors cells to each other within the epidermis so that disruption of the desmosome results in intraepidermal blistering and flaccid bullae. Pemphigus vulgaris often begins with lesions in the oropharynx (recall that this patient had a normal oral exam), which then spread to the skin preferentially affecting the scalp, face, axillae and groin. Nearly all patients have oral lesions at some point during their disease. Skin biopsy shows deposits of IgG between epidermal cells.

B. An antigen in the hair follicle is thought to be the target of autoantibodies in alopecia areata but the precise antigen has yet to be identified.

D. The melanocyte is thought to be the target of autoantibodies in vitiligo.

E. The Pacinian corpuscle is a mechanoreceptor in the skin primarily responsible for sensing deep touch and vibration. It is not a target for autoimmune disease.
A 30-year-old woman presents with chest pain for one day. The pain is in the center of her chest and is worse with deep inspiration and coughing. She reports that it improves when she leans forward while sitting on the examining table. An ECG in the office shows ST segment elevation in all leads. What is the most likely diagnosis?

- A. Aortic dissection
- B. Myocardial infarction
- C. Pancreatitis
- D. Pericarditis
- E. Pulmonary embolism

**Correct answer:** D

This woman’s symptoms and ECG are typical of pericarditis. The other diagnoses may present with similar symptoms, but the ECG findings are diagnostic for pericarditis.

Aortic dissection (A) typically presents with a tearing pain radiating to the back. ST elevation in all leads would not likely be seen.

Myocardial infarction (B) presents with severe substernal pressure-like pain, often radiating to the left arm/jaw. Inspiration, coughing, and positional changes would be unlikely to change the quality of the pain.

Pancreatitis (C) usually presents with severe upper abdominal pain, nausea, vomiting, and anorexia.

Although one of the presenting symptoms of pulmonary embolism (E) can be chest pain that is pleuritic in nature, it is almost always characterized by sudden-onset dyspnea.
An 83-year-old man with a history of exertional dyspnea and lightheadedness experiences a syncopal episode and suffers severe head trauma. Regarding this type of valvular lesion, shown above during postmortem examination, which of the following statements is most correct?

- a. Associated with a harsh diastolic crescendo-decrescendo murmur
- b. Auscultation reveals a paradoxically split second heart sound of decreased intensity
- c. Intensity of murmur predicts severity of disease
- d. Presence of thrill predicts severity of disease
- e. Results in left ventricular dilatation

**Correct answer: B**

The valvular lesion pictured is aortic stenosis (of rheumatic origin in this case; calcific stenosis is also common in this population). In a normal heart, A2 closes before P2; inspiration delays P2 resulting in the physiologically split S2. Aortic stenosis delays A2 until after P2; inspiration thus moves P2 closer to A2, a phenomenon called “reversed splitting.” In addition to this delay in closing, stenosis results in decreased intensity of A2, making choice B correct.

Aortic stenosis is associated with a harsh crescendo-decrescendo murmur during systole, not diastole, therefore choice A is incorrect.

Neither the intensity of murmur (choice C) nor presence of thrill (choice D) predict the severity of disease.

The increased pressures required to overcome aortic stenosis lead to concentric left ventricular hypertrophy. Results in left ventricular dilatation (choice E) is incorrect.
A 55-year-old man presents to the emergency department complaining of sudden-onset tearing chest pain radiating to the back. He denies any history of trauma. Chest radiograph is shown.

What comorbidity is most likely associated with his present condition?

a. Atherosclerosis
b. Churg-Strauss syndrome
c. Hypertension
d. Pericarditis
e. Polycystic kidney disease

Correct answer: C

The radiograph shows a massively enlarged aortic arch, and the given history is classic for a dissecting aortic aneurysm. Hypertension (choice C) is often associated with this pathology. Conditions leading to cystic medial necrosis (e.g. Marfan syndrome) are also associated with aortic aneurysm.

Atherosclerosis (choice A) is incorrect. Atherosclerotic aneurysms usually involve the abdominal aorta, not the aortic arch.

Churg-Strauss syndrome (choice B) is incorrect. This disorder is an allergic granulomatosis typically affecting the lung, spleen, or kidney.

Pericarditis (choice D) is incorrect. Although this condition leads to chest pain, it is typically slower in onset in the absence of trauma, and usually does not radiate to the back. It is often associated with pericardial effusion, not aortic aneurysm.

Polycystic kidney disease (choice E) is incorrect. This disorder is associated with berry aneurysms, typically in the Circle of Willis.
A 49 year old man is found to have a solitary neck nodule on physical examination. Laboratory studies reveal an elevated calcitonin level and fine needle aspiration confirms a diagnosis of medullary carcinoma of the thyroid. Which of the following disease processes might the physician be concerned to look for in this patient?

a. Hyperparathyroidism
b. Papillary thyroid carcinoma
c. Pituitary adenoma
d. Subacute thyroiditis
e. Zollinger-Ellison syndrome

**Correct answer:** A

This man has been diagnosed with medullary carcinoma of the thyroid gland. This is a cancer of the C cells of the thyroid that secretes calcitonin into the bloodstream. Medullary carcinoma can occur as part of the hereditary multiple endocrine neoplasias (MEN) 25% of the time. It can be seen with MEN type II or MEN type III. Hyperparathyroidism can be seen coupled with MENII, aka Sipple syndrome, due to a parathyroid tumor. The other component of Sipple syndrome is pheochromocytoma. MEN III is defined by medullary carcinoma, mucosal neuromas, and pheochromocytoma.

Papillary thyroid carcinoma (choice B) is the most common type of thyroid cancer that is not associated with MEN. A major risk factor for its development is radiation exposure. It is characterized pathologically by psammoma bodies.

Pituitary adenomas (choice C) can be seen in MEN type I, aka Wermer syndrome, in conjunction with pancreatic and parathyroid tumors.

Subacute thyroiditis (choice D) is the most common cause of a painful thyroid gland. It is characterized by a hypothyroid state occurring most often after a flu-like upper respiratory tract infection. It is self limited and not associated with MEN.

Zollinger-Ellison syndrome (ZE syndrome, choice E) is characterized by an increased serum level of gastrin often caused by a gastrinoma of the pancreas or duodenum. Increased gastrin leads to increased hydrochloric acid, predisposing to abdominal pain, diarrhea, and peptic ulcer disease. ZE syndrome, if derived from the pancreas, can be seen in the context of MEN I.
A 33-year-old woman presents complaining of painless rectal bleeding. On three occasions in the last six months, she has noticed a moderate amount of red blood in the toilet after a bowel movement. She denies abdominal pain, constipation, diarrhea, and weight loss. On exam, you notice telangiectasias of her lips and oral mucous membranes. On questioning, she reports that her father also has similar “red spots” on his lips. Rectal exam reveals no mass and no gross blood but she is guaiac positive. What is the most likely source of her bleeding?

a. Arteriovenous malformation
b. Colon cancer
c. Diverticulosis
d. Infectious colitis
e. Ulcerative colitis

correct answer: A

This woman has Osler-Weber-Rendu syndrome (or hereditary hemorrhagic telangiectasia), an autosomal dominant disorder characterized by telangiectasias and arteriovenous malformations (AVMs) of the mucous membranes and the GI tract. A colonic AVM could result in intermittent episodes of rectal bleeding. AVMs are a fairly common cause of GI bleeding, and the majority of patients who have them do not have Osler-Weber-Rendu syndrome. Colon cancer (B) usually presents in an older patient (screening colonoscopy should be started at age 50), unless the patient has familial adenomatous polyposis (FAP) or hereditary nonpolyposis colorectal cancer (HNPCC) - there is no family history in the question stem suggesting either of these. Diverticulosis (C) can cause painless rectal bleeding but would be very rare in a patient this young. Infectious colitis (D) would be associated with diarrhea and crampy abdominal pain, and is inconsistent with the six month time frame. Ulcerative colitis (E) would also be associated with diarrhea and abdominal pain.
A 73-year-old woman has episodes of abdominal pain and increasingly severe constipation. Test of the stool for occult blood is positive. The photomicrograph shows features of the resected colon. Which of the following is the most likely diagnosis?

- a. Adenocarcinoma
- b. Diverticular disease
- c. Polypoid adenoma
- d. Villous adenoma
- e. Volvulus

**Correct answer:** B

The symptoms and histology are typical for diverticulitis, the inflammation of a false pocket (diverticulum) consisting of mucosa and submucosa which have herniated through the muscularis layer. Diverticulitis affects older persons and typically presents with abdominal pain and rectal bleeding. Complications frequently include bowel stenosis, peritonitis, and abscess formation.

Choice A, C and D are incorrect because colon tumors typically present as polyps rather than pockets.

Choice E is incorrect because volvulus, an abnormal twisting of the intestine which impedes blood flow, typically has a sudden onset and does not display the histology shown here.
A 4-year-old girl has the sudden onset of abdominal pain and vomiting. She has a mass in the right lower quadrant and hyperactive bowel sounds. A segment of resected bowel is shown in the photograph. Which of the following is the most likely diagnosis?

- a. Appendicitis
- b. Intussusception
- c. Meckel diverticulum
- d. Necrotizing enterocolitis
- e. Strangulated hernia

**Correct answer: B**

This girl has suffered from intussusception (choice B), when a piece of bowel gets “telescoped” into the proximal bowel, leading to ischemia and infarction. The patient’s abdominal pain and vomiting are consistent with this diagnosis. The picture is key to this question, since all of the other choices could have similar clinical presentations.

Appendicitis (choice A) would also present with RLQ pain and vomiting but would not involve the adjacent bowel.

Meckel diverticulum (choice C) would present similarly to appendicitis but the would show an outpouching of bowel, not an entire infarcted region.

Necrotizing enterocolitis (choice D) is unusual in a four year old; most patients are newborns.

Strangulated hernia (choice E), would present similarly but the pathology would not show one piece of bowel telescoped into the other.
A 21-year-old man has weight loss and severe intermittent bloody diarrhea. A barium enema and colonoscopy show multiple ulcers and inflammatory changes extending from the rectum to the mid-transverse colon. Biopsy specimens taken from multiple sites show acute and chronic inflammation restricted to the mucosa. Which of the following is the most likely diagnosis?

a. AIDS-associated gastroenteritis  
b. Crohn disease  
c. Clostridium difficile-associated colitis  
d. Escherichia coli-associated colitis  
e. Ulcerative colitis

**Correct answer:** H

The key to answering this question is focusing on 4 key points:

1. diarrhea with or without blood  
2. location: inflammatory changes are seen in the **DISTAL** part of the colon only  
3. number of lesions: there are multiple ulcerations  
4. depth of lesion: these ulcerations are limited to the mucosa.

Ulcerative colitis matches the characterization in the question stem along these 4 four points.

The various diseases listed in the answer choices can best be differentiated based on the 4 points listed above. In particular, you must be familiar with the differences between Crohn’s disease (B) and Ulcerative Colitis (E).

(A) AIDS associated gastroenteritis results in pathological changes more proximally than described here, and would not selectively target the colon. Further, AIDS complications rarely result in inflammatory changes.

(B) Crohn's disease may occur anywhere throughout the colon and occurs in a patchy, discontinuous fashion. It penetrates deeper into the mucosa producing transmural fistulae and inflammation, unlike UC.

(C) C.difficile colitis (also called pseudomembranous colitis) typically affects older patients who have had prior exposure to antibiotics and rarely produces bloody diarrhea.

(D) E. coli is the most common cause of bacterial colitis, and depending on the pathogenic strain, may present differently. Notably, the lamina propria of the large intestine is infiltrated by polymorphonuclear leukocytes, without producing such a pattern of mucosal inflammation.
A 35-year-old woman presents with worsening right upper quadrant pain for two days. For the past 3 months she’s had a similar pain 1-2 hours after meals, but now the pain is more severe and constant. She has also felt feverish and complains of clay-colored stools, dark urine, and a yellowish skin color. What is the most likely diagnosis?

- a. Acute cholecystitis
- b. Ascending cholangitis
- c. Biliary colic
- d. Choledocholithiasis
- e. Pancreatitis

**Correct answer:** B

Ascending cholangitis is thought to be an ascending bacterial infection related to (D) choledocholithiasis and bile stasis. Charcot’s triad of RUQ pain, fever, and jaundice are typical of ascending cholangitis. Reynold’s pentad includes hypotension and altered mental status in addition to Charcot’s triad.

(A) The presence of clay-colored stools, dark urine, and jaundice suggests that the blockage is distal to the cystic duct (e.g. in the common bile duct), making acute cholecystitis incorrect.

(C) Biliary colic does not present with fever, jaundice, dark urine, and clay-colored stools.

(D) Uncomplicated choledocholithiasis does not present with fever and intense pain.

(E) Gallstones in the common bile duct can cause pancreatitis, but pancreatitis is unlikely to be the primary illness in this case.
A 65-year-old man from China presents to his doctor complaining of increased abdominal girth, weight loss, and fever. Physical exam is remarkable for right upper quadrant tenderness and organomegaly. Tissue biopsy is positive for a primary malignancy.

What tumor marker is most likely elevated in this patient?
   a. Alphafetoprotein (AFP)
   b. CA-125
   c. CA19-9
   d. Carcinoembryonic antigen (CEA)
   e. S-100

*correct answer: A*

This patient’s symptoms are consistent with a hepatic process. Of note, China is an area of endemic Hepatitis B infection, a risk factor for hepatocellular carcinoma (HCC). Alphafetoprotein (AFP) (choice A) is associated with HCC and nonseminomatous testicular germ cell tumors.

CA-125 (choice B) is associated with ovarian cancer.

CA19-9 (choice C) is associated with pancreatic cancer.

Carcinoembryonic antigen (CEA) (choice D) is associated with carcinomas of the lung, pancreas, stomach, breast, and colon. Though colon cancer commonly metastasizes to the liver, biopsy in this patient indicates a primary, not metastatic, malignancy.

S-100 (choice E) is associated with melanoma and neural-derived tumors.
An otherwise healthy 26-year-old woman has had petechiae on her legs during the last 24 hours. Laboratory studies results are above. A peripheral blood smear shows normal red cell morphology; a bone marrow smear shows mature megakaryocytic hyperplasia. Which of the following is the most likely diagnosis?

a. Acute megakaryocytic leukemia
b. Acute myelogenous leukemia
c. Aplastic anemia
d. Immune thrombocytopenic purpura
e. Epstein-Barr viral infection

**Correct answer:** D

Immune thrombocytopenic purpura (ITP) (D) causes a profound decrease in platelets, can reveal petechiae on physical exam, and should be of rapid onset with all other laboratory values normal. Bone marrow biopsy would reveal normal-to-increased numbers of megakaryocytes.

Marrow biopsy in acute megakaryocytic leukemia (A) would show hypercellular marrow replaced by blast cells and dysmorphic promegakaryocytes and megakaryocytes. This patient has mature megakaryocytes.

In acute myelogenous leukemia (B), blast red cells should be seen in the peripheral smear and bone marrow. This patient has normal red cell morphology.

In aplastic anemia (C) there are profound decreases in red cells, white cells, and platelets. This patient has normal RBC and WBC values.

Epstein-Barr (EBV) viral infection (E) is usually associated with a leukocytosis and mild thrombocytopenia, not the normal WBC count and profound platelet deficiency seen here. Though immune thrombocytopenic purpura is a rare complication of EBV, (D) is a better answer choice in this situation.
You are examining a patient’s peripheral blood smear under a light microscope. The patient is a 25-year-old woman who complains of increasing fatigue over the last several months. She tells you her mother and sister had similar complaints when they were in their twenties. She also reports having heavy menses.

The smear is notable for considerable variation in the size and shapes of the RBCs, though many appear smaller than normal. You don’t notice any schistocytes.

From these observations, what is the most likely condition the patient has?

a. Hereditary spherocytosis
b. Iron-deficiency anemia
c. Megaloblastic anemia
d. Sickle cell anemia
e. Thalassemia

_correct answer: B_  
This patient likely has iron-deficiency anemia secondary to heavy menses. The cells appear smaller, which is inconsistent with megaloblastic anemia (where the cells, and MCV are larger). The description of the peripheral smear includes variation in cell size (anisocytosis) and cell shape (poikilocytosis), which are characteristic of iron-deficiency anemia. This is in contrast to thalassemia, where the cells are more uniformly small.

Hereditary spherocytosis could present similarly, but is less common and is more likely to have presented earlier with anemia, icterus/jaundice, and/or splenomegaly. Sickle cell anemia would likely have been caught at birth, and the patient's presentation is not consistent with this severe disorder.
A 41-year-old man presents to clinic complaining of vague abdominal pain and fatigue. History is remarkable for several episodes of pneumonia over the past year. Physical examination reveals marked splenomegaly. CBC shows a hemoglobin concentration of 8.2 g/dL, hematocrit of 24.6%, MCV of 90 fL, WBC count of 2400 and platelet count of 63,000.

Peripheral blood smear demonstrates many small leukocytes with kidney-shaped nuclei and pale blue cytoplasm with threadlike extensions. Immunohistochemical staining is positive for CD20.

Which of the following additional findings is most characteristic of this disease?

a. Auer rods in leukocytes  
b. Monoclonal IgM spike in serum  
c. Philadelphia (Ph) chromosome expression  
d. Tartrate-resistant acid phosphatase in leukocytes  
e. Toxic granulations in neutrophils

**Correct answer:** D

This patient has hairy cell leukemia, an uncommon neoplastic disorder of B cells (CD20+). These cells infiltrate the spleen and marrow. Pancytopenia results from poor production of hematopoietic cells in the marrow and sequestration of the mature cells in the spleen. There are two characteristic findings with this disease: hairy projections from neoplastic leukocytes in the peripheral blood smear, and tartrate-resistant acid phosphatase in leukocytes (Choice D).

Auer rods (Choice A) are characteristic of myeloblasts in acute myelogenous leukemia (AML).

Philadelphia (Ph) chromosome expression (Choice C) is a classic finding for chronic myelogenous leukemia (CML), but is occasionally seen in acute lymphoblastic leukemia (ALL) and acute myelogenous leukemia (AML).

Toxic granulations in neutrophils (Choice E) are typically seen in overwhelming bacterial infections.

A monoclonal IgM spike is a feature of lymphoplasmacytic lymphoma such as Waldenstrom macroglobulinemia.
While skipping rope, a 7 year old girl falls and scrapes her knee on the pavement. Four days later, she picks at a hard, reddish-brown raised formation at the site of the injury which reveals soft, moist pink tissue. Which is the predominant cell type found in this underlying tissue?

- a. Epithelioid macrophage
- b. Fibroblast
- c. Mast cell
- d. Neutrophil
- e. Platelet

correct answer: B

Granulation tissue forms within 3-5 days after severe or persistent injury has occurred. It is characterized by proliferation of fibroblasts and new capillaries that will form the scaffolding for scar formation. Epithelioid macrophages are large and squamous-cell like, found in chronic inflammation, such as in tuberculosis or sarcoidosis. Neutrophils are the predominant leukocytes found in acute inflammation, usually within the first 24 hours of injury. Platelets create a "plug" at the site of bleeding on which a fibrin clot forms a scab within the first few hours of injury. Mast cells release histamine during acute inflammation, which causes vasodilation and increased vascular permeability.
A 42-year-old woman felt a mass in her left breast while taking a shower. She was later diagnosed with infiltrating ductal carcinoma. The mass is removed with lumpectomy along with an axillary lymph node dissection. Which of the following findings will best predict a better prognosis for this patient?

a. Aneuploidy and high S-phase levels found via flow cytometry
b. Estrogen receptor not expressed on tumor cells
c. Family history of breast cancer
d. Histological findings of high grade tumor
e. Lymph node biopsies are negative

**Correct answer:** E

Lymph nodes without any tumor cells are a good predictor of better survival. It is an indication of metastasis free breast cancer.

All other choices are associated with bad prognosis. High grade tumors are very poorly differentiated and does not resemble breast tissue. Family history of breast cancer is bad. ER- is associated with poor prognosis. High S-phase means the tumor cells are dividing at a much higher rate.
A patient with malignant lymphoma is treated with a chemotherapeutic agent that results in nuclear and cytoplasmic fragmentation of individual cells. Which of the following mechanisms is the most likely cause of the cellular fragmentation?

a. Acute inflammation
b. Apoptosis
c. Autophagy
d. Mitochondrial poisoning
e. Necrosis

_correct answer: B_

Apoptosis is the process that is described here. Along with cellular fragmentation, DNA laddering and cytochrome c release are also seen.

The other choices do not result in cell fragmentation.
A 5 year old boy's height is found to be in the lowest 5th percentile for his age. He receives minimal sunlight during the day and he is lactose intolerant. He otherwise has no known health problems and takes no medications/supplements. Which of the following most likely describes X-ray findings of his hips and lower extremities?

- a. Diffuse osteolytic lesions filled with cystic spaces
- b. Diffuse radiolucency with excess osteoid and bowing
- c. Diffusely increased bone density
- d. Normal lucency, architecture and posture
- e. Patchy blastic and lytic lesions throughout pelvis and femurs

**Correct answer:** B

The child has signs of Vitamin D deficiency, or rickets, secondary to inadequate sunlight exposure and poor calcium intake. Vitamin D is necessary to absorb calcium and phosphate from intestinal mucosa. In its absence, bone cannot be mineralized, so there is an excess of osteoid (unmineralized bone) and diffuse osteopenia. Bowing, a.k.a. genu varum, is a deformity of the legs that characterizes rickets. In adults this condition is called osteomalacia.

Diffuse osteolytic lesions filled with cystic spaces describes osteitis fibrosa cystica, an osteolytic condition secondary to hyperparathyroidism where resorbed bone is replaced by fibrous tissue. Patchy blastic and lytic lesions throughout pelvis and femurs characterizes Paget's disease, a disorder of deranged bone remodeling that usually affects the elderly. Diffusely increased bone density occurs in Osteopetrosis, which is caused by osteoclast dysfunction leading to susceptibility to fractures from "marble bones." Without Vitamin D supplementation and adequate calcium intake, growing children have no source of minerals to build bone, and can develop permanent skeletal deformities.
A 71-year-old man presents to his primary care provider complaining of nonradiating lower back pain over the past several months. On review of symptoms, he mentions feeling shorter than previously. Physical exam reveals a slight abnormal curve of the spine. Radiographic studies show anterior vertebral wedging and vertebral end-plate irregularity.

What is the most likely underlying pathology of this process?

- Decreased osteoblast activity, normal osteoclast activity
- Increased osteoblast activity, decreased osteoclast activity
- Intervertebral disc degeneration
- Intervertebral disc space calcification
- Decreased osteoblast activity, increased osteoclast activity

**Correct answer:** A

This patient is suffering from a compression fracture secondary to osteoporosis. Decreased osteoblast activity with normal osteoclast activity (choice A) will result in an overall decrease in bone density. This process occurs in primary osteoporosis type 2 (“senile osteoporosis”), which occurs in patients over 60 of both genders.

Increased osteoblast activity with decreased osteoclast activity (choice B) will result in an overall increase in bone density. This process is not associated with vertebral compression fractures.

Intervertebral disc degeneration (choice C) leads to disc herniation. This condition can cause lower back pain secondary to nerve root compression, classically radiating to the upper legs (“sciatica”). This pathology is inconsistent with the observed radiographic studies.

Intervertebral disc space calcification (choice D) typically affects preadolescent children. It usually is found in the thoracic and lumbar spine, and the course is self-limited.

Decreased osteoblast activity with increased osteoclast activity (choice E) will result in an overall decrease in bone density and may lead to a vertebral compression fracture. This process occurs in primary osteoporosis type 1 (“postmenopausal osteoporosis”), which is six times more common in women. In this process, loss of estrogen is thought to increase cytokine levels promoting osteoclast activity.
A 16-year-old African-American boy presents to his primary care physician complaining of leg swelling for the past several months. Exam reveals a large, tender soft tissue mass just proximal to the knee. Of note, he is found to have lost 8 lbs and has a hemoglobin level of 11.0 g/dL. Plain film demonstrates periosteal elevation with bone destruction; a biopsy is performed.

Which of the following describes the expected pathology specimen?
- a. Brown nodule surrounded by dense sclerotic cortical bone
- b. Dense normal bone with cartilaginous cap
- c. Large, necrotic, hemorrhagic mass with osteoid and bone
- d. Myxomatous tissue
- e. Neoplastic colon tissue

**correct answer:** C

With regards to race, age, gender, complaint, duration, and location, this patient demonstrates a classic presentation the most common primary bone tumor: osteosarcoma. (Systemic symptoms are variable.) Presence of osteoid is the defining characteristic for all types of osteosarcoma. Large, necrotic, hemorrhagic mass with osteoid and bone (choice C) is correct.

Brown nodule surrounded by dense sclerotic cortical bone (choice A) is seen with osteoid osteoma. This condition would not demonstrate the radiological findings described.

Dense normal bone with cartilaginous cap (choice B) describes an exostosis of a benign osteochondroma. This growth would be visible on x-ray, and does not lead to periosteal elevation or bone destruction.

Myxomatous tissue (choice D) is seen in lesions of chondromyxoid fibroma; however, these lesions are usually intramedullary rather than cortical.

Neoplastic colon tissue (choice E) could be seen with metastatic colon cancer, but is unlikely in this patient due to his age. Further, among metastatic cancers, colon is least likely to metastasize to bone.
A 26-year-old medical student falls while snowboarding and strikes the left side of his head against the hard packed mountain slope. He was not wearing a helmet. A head CT scan reveals a convex, lens-shaped area of hemorrhage centered over the left parietal region. Which of the following intracranial vascular structures is likely responsible for this bleeding?

a. Cavernous sinus  
b. External carotid artery  
c. Inferior cerebellar artery  
d. Left parietal artery  
e. Middle meningeal artery

**correct answer:** E

Damage to the middle meningeal artery is the cause of an epidural hematoma. In this case, it is secondary to blunt trauma of the left temporal region. Epidural hematomas are characterized by convex, lens-shaped areas of hemorrhage near the parietal bone.
A 45-year-old man has abnormal circadian variation in body temperature, disruption of the sleep-wake cycle, and an impaired nocturnal surge of secretion of melatonin. An MRI of the brain is most likely to show a lesion involving which of the following nuclei?

- a. Accessory optic
- b. Lateral preoptic
- c. Pretectal
- d. Suprachiasmatic
- e. Supraoptic

**Correct answer:** D

The nucleus involved with circadian rhythms, sleep-wake cycle and melatonin regulation is the suprachiasmatic nucleus of the hypothalamus (option D).

Accessory optic nucleus (A) is involved in eye movements.

The lateral preoptic nucleus (B) is part of heat regulation in the hypothalamus.

The pretectal (C) is the area in the midbrain related to pupillary light reflex.

The supraoptic nucleus (E) of the hypothalamus regulates water balance and the produces ADH and oxytocin.
A 50-year-old man is brought to the emergency room complaining of a severe headache. The headache began 15 minutes earlier and has not subsided. There is no recent history of head trauma and his vision is normal.

His blood pressure is 110/60. His heart rate is 85 bpm. Upon lumbar puncture, his sampled CSF appears bloody.

Which of the following is most likely underlying cause of this man’s distress?

a. Epidural hematoma  
b. Parenchymal hematoma  
c. Subarachnoid hemorrhage  
d. Subdural hematoma  
e. Temporal arteritis

Correct answer: C

A subarachnoid hemorrhage (C) results from the rupture of an aneurysm (usually berry) or an arteriovenous malformation (AVM). Spinal tap reveals a bloody or xanthochromatic CSF specimen.

Epidural (A) or subdural (D) hematomas most likely result from blunt trauma to the head. While epidural hematomas are often immediate and secondary to fracture of the temporal bone, the subdural variety are insidious and a delayed consequence of bridging vein rupture.

Parenchymal hematomas (B) are caused by hypertension, amyloid angiopathy, diabetes mellitus and tumors. There is no such history here to support this diagnoses.

Temporal arteritis (E) is the most common vasculitis and affects arteries from large to small size. Symptoms are not as rapid in onset as described here and include jaw claudication and impaired vision.
A 25 year old woman presents to the ED with drooling out of the right side of her mouth. She says that she first noticed the drooling in the morning when she woke up. Further questioning reveals that she has had some ear pain for the past few days. Physical exam reveals small, red vesicles in the right external meatus. Which of the following additional symptoms may be found in this patient.

a. Ophthalmoplegia of the right eye on abduction
b. Hyperacusis
c. Presbycusis
d. Voice hoarseness
e. Anosmia

correct answer: B

This patient is most likely suffering from a Bell’s palsy secondary to herpes zoster flare. Bell’s palsy is classically described as paralysis of facial muscles due to trauma/inflammation of the facial nerve, CN VII. There are a few etiologies of Bell’s palsy which include various herpes viral infections, Lyme disease, HIV, and sarcoidosis. In addition to facial muscle paralysis, there are other symptoms associated with damage to the facial nerve. The facial nerve is a mixed nerve that contains fibers to salivary glands, afferent fibers for taste, and efferent fibers to the stapedius muscle in the middle ear. The stapedius muscle functions to control the amplitude of sound waves transmitted to the inner ear by stabilizing the stapes. Paralysis of the stapedius will result in hyperacusis (choice B) in which normal sounds are perceived to be louder.

The inability to abduct the right eye (choice A) suggests paralysis of the lateral rectus muscle which is innervated by CN VI. CN VI should not be affected in CN VII inflammation.

Presbycusis (choice C) is progressive hearing loss usually found in the elderly. High-frequency sounds are preferentially lost first.

Voice hoarseness (choice D) can be observed in unilateral damage to the recurrent laryngeal nerve. The RLN is a branch of the vagus nerve, CNX, that carries fibers from the spinal accessory nerve, CNXI.

Anosmia (choice E) is the loss of olfactory sense. Although facial nerve inflammation can affect sense of taste, sense of smell, CN I, in unlikely to be affected.
A 60 year old man with no significant family or past medical history has had a significant decline in his cognitive abilities in the past 3 months. His initial diagnostic testing was unremarkable with normal blood values and imaging. He continues to deteriorate and develops a right hemiparesis. 1 month later the patient develops aspiration pneumonia and expires. Autopsy reveals the following finding on brain microscopy. What is the protein most likely responsible for this patient's condition?

- a. β-amyloid
- b. Hyperphosphorylated tau protein
- c. Ubiquinated neurofilaments
- d. Proteinase-resistant protein
- e. Negri bodies

**Correct answer:** D

This patient has Creutzfeld-Jakob disease which is a spongiform encephalopathy that produces rapidly progressive dementia over the course of months. Brain microscopy is characterized by a “bubbles and holes” pattern which is exemplified in the histological image. The disease is caused by accumulation of an abnormal protein that resists degradation by proteinases; hence, the protein is called proteinase-resistant protein (choice D) or PrP or prion protein.

β-amyloid (choice A) and hyperphosphorylated tau protein (choice B) are found in Alzheimer’s disease. AD is the most common cause of dementia; however, the developmental time course is much slower than described in this patient and there would be senile plaques on histological studies.

Ubiquinated neurofilaments (choice C) are the protein basis for Lewy bodies found in Parkinson’s disease. Parkinson’s is also a cause of dementia; however, it is also commonly associated with a resting tremor and cogwheel rigidity which is not described in this patient.

Negri bodies (choice E) are eosinophilic, intracellular inclusion bodies found in Purkinje cells that are pathognomonic for rabies infection. These bodies are not protein, but are aggregates of rabies virus.
A 6-year-old girl has the sudden onset of swelling of her face, hands, legs, and feet 1 week after a viral upper respiratory tract infection. She is afebrile and normotensive. Laboratory studies show:

- Serum Albumin 2.0 g/dL
- Serum Urea nitrogen 6.0 mg/dL
- Serum Creatinine 0.6 mg/dL
- Serum Cholesterol 280 mg/dL
- Urine protein 4+; 6.0 g/24 h

**Which of the following is the most likely diagnosis?**

a. Focal glomerulosclerosis  
b. Membranous glomerulonephritis  
c. Membranoproliferative glomerulonephritis  
d. Minimal change disease  
e. Rapidly progressive glomerulonephritis

**correct answer:** D

The sudden onset of edema in the face and dependent limbs, coupled with hypoalbuminemia suggests a nephrotic syndrome in this child. Consistently, she has hyperlipidemia (cholesterol >200mg) and proteinuria. In children, the most common cause of a nephrotic syndrome is Minimal Change Disease (D, abbrev. MCD).

The child does not exhibit hypertension or overt hematuria. This eliminates renal diseases that are associated with a more NEPHRITIC picture, such as membranoproliferative glomerulonephritis (C) and Rapidly Progressive GN (E).

Be sure to review NEPHROTIC vs NEPHRITIC syndromes.

MCD, (aka lipid nephrosis) involves T cell mediated destruction of the polyanion charge barrier on epithelial foot processes, which results in leakage of albumin into the urine. This is manifest in this patient as a reduced serum Albumin (<3.5mg) and Proteinuria (>0.15g/24h). While MCD is often idiopathic, it has been described to occur days to weeks after upper respiratory infections, as here.

It is difficult to differentiate between MCD and the other nephrotic choices clinically, but you should know these important distinctions in their histology:

By definition, patients with MCD present histologically with normal looking glomeruli.

Focal segmental Glomerulosclerosis (A, FSGS) presents with focal sclerotic plaques of the glomerular tufts and hyaline deposits. It is also often associated with HIV and IV drug use.

Membranous glomerulonephritis (B) presents later in life, insidiously and is an immune complex disorder that results in mesangial deposits. The workup will show positive Anti nuclear antibodies and/or Anti dsDNA. (remember, this is a NEPHROPATHY and is assoc. with a NEPHROTIC picture despite its name).
A 4 year old boy is brought in by his mother after she discovers that his urine is now a dark coca-cola color. She reports that he was at the doctor 10 days ago because he was complaining of a sore throat, but he has otherwise been well. What is the most likely diagnosis?

a. Alport's syndrome
b. Goodpasture's syndrome
c. Henoch-Schonlein purpura
d. Minimal change disease
e. Post-streptococcal glomerulonephritis

**Correct answer:** E

The history of possible streptococcal pharyngitis and lack of other symptoms makes post-streptococcal GN the most likely answer.

(B) Goodpasture's does not typically present before young adulthood and results in destruction of pulmonary and glomerular basement membranes.

(C) HSP is a vasculitic syndrome that often includes purpura, abdominal pain, and arthritis. Glomerulonephritis is often seen.

(D) Minimal change disease is a nephrotic syndrome. The primary complaint of dark colored urine and recent possible streptococcal infection makes a nephritic syndrome more likely.
Breast carcinomas often cause the skin of the breast to become puffy and pitted, resembling orange peel. The pits most likely correspond with which of the following?

- a. Attachments of suspensory ligaments (retinacula cutis) to the dermis
- b. Diffuse scarring in subcutaneous fibrous tissue
- c. Focal invasion of the dermis by neoplastic cells
- d. Openings of sebaceous gland
- e. Openings of sweat glands

**correct answer:** A

Inflammatory carcinoma of the breast creates pitting classically referred to as an "orange peel" appearance on clinical exam. It produces a rapid increase in breast size, itching, redness and warmth of the overlying skin. There is usually no obvious lump, and likewise inflammatory carcinoma is often confused with mastitis on clinical exam. The pitting is caused by severe inflammation destroying the suspensory (Cooper's) ligaments attaching breast tissue to the dermis, resulting in a dimpled "orange peel." The remaining answers do not play a part in the pathogenesis of pitting in inflammatory carcinoma.
A 55-year-old woman has a hysterectomy and bilateral salpingo-oophorectomy for abnormal uterine bleeding. Histologic examination of the ovaries shows small atretic follicles. Which of the following processes is the most likely cause of these histologic findings?

a. Apoptosis  
b. Metamorphosis  
c. Metaplasia  
d. Necrosis  
e. Transformation

**Correct answer:** A

Small, atretic ovarian follicles are a normal finding in post-menopausal women. As women enter menopause, their ovaries lose the ability to support viable ova and follicles. This process of planned cell death is called apoptosis. By contrast, necrosis is unplanned cell death caused by an insult to previously viable tissue. Metaplasia and transformation are associated with neoplastic processes, which may be occurring in this woman's uterus leading to bleeding. Don't be fooled, the question asks not about uterine histological findings, but rather about those in the ovaries which are entirely benign.
A 34-year-old nulligravid woman comes to clinic complaining of difficulty getting pregnant for the past 15 months. The woman reports that her menses have always been irregular. The woman has a BMI of 31 and mild hirsutism on physical exam. What is the most likely cause of infertility in this patient?

a. Anovulation  

b. Antiphospholipid syndrome  

c. Endometriosis  

d. Submucosal uterine leiomyomas  

e. Unexplained infertility

**correct answer:** A

The correct answer is anovulation. This woman's history is significant for irregular menses, which is often a sign of anovulation. In addition, she is obese with a BMI of 31 and mild hirsutism. This woman likely has polycystic ovarian syndrome leading to ovulatory dysfunction and hyperandrogenism and her infertility is secondary to the anovulation. The next step in the workup would be a progesterone challenge test and if positive, this would confirm anovulation.

The other choices are all causes of infertility that are less likely given this history and physical findings. Patients with antiphospholipid syndrome often present with multiple spontaneous abortions in the first trimester. This patient has never been able to get pregnant so this syndrome is a less likely reason for her infertility.

Endometriosis is a common cause of infertility. However, patients often present with severe dysmenorrhea. Since there is no mention of dysmenorrhea, this etiology is less likely.

A submucosal uterine leiomyoma most likely would present with heavy bleeding and if the patient were ovulating regularly, regular menstrual cycles. There is no mention of heavy or increased bleeding for this patient and given the facts of the case, anovulation is more likely.

If this patient was ovulating regularly and additional tests were unrevealing, then this patient would be diagnosed with unexplained infertility. However, given the constellation of findings, the most likely diagnosis is anovulation secondary to PCOS.
A 44 year-old obese woman who has been recovering from a knee sprain develops sudden shortness of breath and left-sided chest pain with inspiration. On exam, HR is 110, RR is 24, Temp is 37 C, the pulmonic portion of the second heart sound is accentuated, 3 cm of JVD is appreciated, and the right leg is slightly swollen and tender. Trachea is midline and lung sounds are equal bilaterally without wheezes, rales, or rhonchi. The remainder of physical exam is unremarkable. If pathological examination was performed, what would be seen in the affected lung tissue?

a. Caseating necrosis with acid-fast bacilli
b. Exudative consolidation with numerous Gram-positive cocci
c. Hemorrhagic wedge-shaped infarct
d. Loss of alveolar septa
e. Pale infarct at the periphery of the lung parenchyma

**Correct answer:** C

This patient has a pulmonary embolus (PE). This patient is at known risk due to her obesity and immobility (suggested by recent knee sprain). The sudden onset, pleuritic chest pain, and findings on exam are consistent with PE. Since both pulmonary arteries and bronchial arteries supply the lungs, a hemorrhagic (red) infarct would be seen with occlusion of a pulmonary arterial branch. The wedge shape represents the area of lung tissue normally supplied by that branch. Sudden onset and lack of fever make tuberculosis (caseating necrosis with acid-fast bacilli) or bacterial pneumonia (exudative consolidation with Gram-positive cocci) far less likely. Loss of alveolar septa is seen in emphysema.
A 64 year-old shipbuilder arrives at your office complaining of six months of worsening exertional dyspnea. He is a lifelong non-smoker. A chest radiograph reveals pleural thickening. What other finding would be expected in this patient?

a. Acid-fast bacilli in bronchial mucous  
b. Crackles at the lung apices  
c. Decreased FEV1/FVC ratio  
d. Ferruginous bodies on lung biopsy  
e. Progressive cough

**correct answer:** D

The patient must have a history of asbestos exposure as evidenced by his occupational history and the pathognomonic finding of pleural plaques (thickening). The Ferruginous bodies found in his lungs are composed of asbestos fibers coated with iron and proteins. While not exclusive to this pneumoconiosis, they are frequently found on tissue biopsy. Asbestos exposure tends to cause an interstitial lung disease that is classically worse at the lung bases.

Although smoking frequently happens to be comorbid with asbestosis, the question stem clearly states that this patient has never smoked. Progressive cough, and a decreased FEV1/FVC ratio are hallmarks of COPD, but interestingly are unlikely to be found in isolated asbestosis. There is no reason to believe that this patient has active tuberculosis, so finding AFB would be rather unexpected.
A 4 year old girl presents with runny nose, mild fever, distinct "barking cough" and inspiratory stridor. Which of the following conditions is she most likely to have?

- a. Bronchiectasis
- b. Bronchiolitis
- c. Bronchitis
- d. Laryngotracheitis
- e. Pneumonia

**Correct answer: D**

Infectious agents in the lungs tend to have predilections for particular locations in the respiratory system. Acute laryngotracheitis, or croup, is caused by respiratory viruses such as parainfluenza and most commonly affects young children. Stridor occurs because the inflamed vocal cords collapse with the negative pressure of inspiration. Bronchitis affects the large airways, impairing normal ciliary function and causing productive cough. Bronchiolitis is caused by respiratory viruses such as RSV, affects smaller airways, and rarely affects patients older than age 2. Bronchiectasis is bronchial dilation and mucus plugging that occurs after chronic inflammation, such as in cystic fibrosis or recurrent pneumonia. Pneumonia is infection of the alveoli, which typically causes productive cough.
A 34-year-old woman has had a nonproductive cough for 2 months. She has never smoked. An x-ray of the chest shows bilateral interstitial markings and hilar adenopathy. Which of the following findings is most likely on transbronchial biopsy?

- a. Areas of hemorrhagic infiltrate
- b. Areas of liquefaction necrosis
- c. Dilation of respiratory bronchioles
- d. Microabscesses
- e. Noncaseating granulomas

**Correct answer:** E

This woman’s clinical and radiographic history is consistent with the diagnosis of sarcoidosis. Sarcoidosis is characterized by widespread noncaseating granulomas, elevated serum ACE levels, and is associated with, among other things, restrictive lung disease.

(A) Pulmonary hemorrhage is unlikely. There is no history of hemoptysis or evidence of hemorrhage on chest x-ray.

(B) Liquefactive necrosis is often associated with cellular destruction, often secondary acute infection.

(C) There is no suggestion of bronchiectasis or dilation of the respiratory bronchioles on history or radiography.

(D) Bacterial infection with abscess formation is unlikely given this woman’s clinical history and radiographic findings.
Scanning of inhaled, radioactively labeled aerosols has been used to study the distribution of pulmonary ventilation. However, data obtained using aerosols with a particle diameter of 20 micrometers do not accurately reflect the distribution of alveolar ventilation in the lungs. Which of the following is the best explanation for this finding?

a. Ingestion of these particles by alveolar macrophages occurs nonuniformly
b. The majority of particles this size do not adhere to airway or alveolar walls
c. Many particles are rapidly absorbed into the pulmonary capillary blood
d. Most particles this size are deposited in the conducting airways
e. Removal of particles by the mucociliary system is too fast to allow scanning

**Correct answer:** D

The test is attempting to measure the alveolar ventilation, or the amount of gas reaching the alveoli that is capable of gas exchange. The 20 micrometer particles are not arriving as they are too large to pass through the 10+ levels of conducting airways.

One can reach this answer through elimination of the others:
A - macrophage ingestion takes a long time, and would not affect a quick scan such as this. There is no evidence that nonuniform ingestion, if it exists, can explain the problem with this aerosol.

B - If the particles adhered to the walls, the more distal parts of the lung would be less ventilated by the particles, which would exacerbate the problem with the aerosol.

C - This would happen uniformly throughout the lung, which would not affect the measurement of alveolar ventilation.

E - The mucociliary system is exceedingly slow - it is an "elevator." This alone cannot explain the difficulty with the aerosol.
A 41 year old female comes to your office with a complaint of blurry vision over the past one year. On physical exam, you note keratoconjunctivitis and atrophy of the oral mucosa. You decide to biopsy a section of her lip which shows lymphocytic and plasma cell infiltrates in the salivary glands. Which of the following autoantibodies is most likely to be identified on laboratory testing?

a. Anticentromere 

b. SS-B 

c. Jo-1 

d. Anti-double stranded DNA 

e. Scl-70 

Correct answer: B

This question requires you to both identify the disease process described in the stem and then identify a commonly associated antibody used for diagnosis. This stem describes a classic presentation of a patient with Sjogren's syndrome which is caused by an immune mediated attack on the salivary and lacrimal glands. Classic symptoms include dry eyes, dry mouth, dental caries, dysphagia and hoarseness. SS-BB antibodies are found in the majority of patient's with Sjogren's syndrome and are a commonly tested association on board exams. Anticentromere (option A) is most commonly associated with limited scleroderma (CREST syndrome). Jo-1 (option C) is a marker for polymyositis. Anti-dsDNA (option D) is an autoantibody specific for systemic lupus erythematos, and Scl-70 (option E) a marker for diffuse systemic scleroderma.
An 82 year-old male presents to his physician with complaint of leg swelling. He was diagnosed with prostate cancer two years ago. What is the most likely cause of the patient's edema?

- a. Decreased capillary hydrostatic pressure
- b. Decreased interstitial osmotic pressure
- c. Increased capillary osmotic pressure
- d. Increased capillary permeability
- e. Occlusion of lymph vessels

**Correct answer:** E

Pressure\_net = \text{Capillary permeability} \times [(\text{Pressure capillary} + \text{Osmotic capillary}) - (\text{Pressure interstitial} + \text{Osmotic interstitial})]. A, B, and C all result in a net fluid flow into the capillary. Increased capillary permeability is bi-directional, so it won't result in pooling of fluid. However, occlusion of the lymph vessels results in interstitial pooling. The prostate cancer diagnosis is a hint to metastatic cancer into the lymph vessels.
A 60 year-old man with coronary artery disease has been hospitalized for a large myocardial infarction he suffered 5 days ago. He suddenly develops hypotension and prominent jugular vein distention. On cardiac exam, his heart sounds are muffled. His pulse is palpably weaker during inspiration. What is the most likely diagnosis?

a. Arrhythmia  
b. Myocardial infarction  
c. Myocardial wall rupture  
d. Pericarditis  
e. Ruptured papillary muscle

**Correct answer:** C

This patient has developed cardiac tamponade (acute build-up of fluid in the pericardial space) 5 days after a myocardial infarction. This patient has all three features of Beck’s triad: hypotension, JVD, and distant heart sounds. Myocardial wall rupture most commonly occurs between 4 and 7 days after an MI.
A 28-year-old man who had rheumatic fever as a child comes to the physician’s office because of fatigue and dyspnea for the past 4 months. An early diastolic sound followed by a low-pitched rumbling decrescendo diastolic murmur is present 4 cm left of the sternal border in the fourth intercostal space and is heard best with the patient in the left lateral decubitus position. Which of the following valve defects is most likely in this patient?

a. Aortic regurgitation  
b. Aortic stenosis  
c. Mitral regurgitation  
d. Mitral stenosis  
e. Pulmonic regurgitation

**Correct answer:** D  
Always associate mitral stenosis with rheumatic fever. Other valves can be involved but the most common cause of mitral stenosis is aseptic scarring after systemic Group A Streptococcus infection. The murmur of mitral stenosis is described as above. As with any murmur, as the severity increases so does pressure in the chamber before the valve (in this case the left atrium). Dyspnea, fatigue, and rales on exam are symptoms of congestive heart failure, regardless of the underlying cause. This is also the murmur that may be revealed during pregnancy. Aortic regurgitation is a diastolic murmur characterized by wide pulse pressure and may be associated with aortic dissection or trauma. Aortic stenosis usually occurs in elderly patients secondary to calcifications, but it can also be secondary to congenital bicuspid aortic valve in young people. The aortic stenosis murmur is crescendo decrescendo and radiates to the carotids. Mitral regurgitation murmurs are systolic and can occur secondary to chordae tendinae rupture after MI or in predisposed people with mitral valve prolapse. It is a blowing mid to late systolic murmur. Pulmonic regurgitation is a diastolic murmur best heard in the left 2nd intercostal space usually secondary to congenital defect.
A 72-year-old man collapses while playing golf. He has a 5-year history of angina and type 2 diabetes mellitus. Paramedics arrive in 10 minutes. Examination shows no respirations or blood pressure; an ECG shows asystole. Cardiopulmonary resuscitation is attempted for 10 minutes without success. Which of the following is the most likely cause of death in this patient?

- a. Cardiac tamponade
- b. Embolus to the right middle cerebral artery
- c. Necrosis of the myocardium
- d. Rupture of the papillary muscle
- e. Ventricular fibrillation

correct answer: E

This man likely had ventricular fibrillation secondary to ischemic heart disease given his age, history of angina and type 2 diabetes, and his sudden collapse and death. Ventricular fibrillation is the most common cause of sudden cardiac death in adults.

(A) Cardiac tamponade, (C) necrosis of the myocardium, and (D) rupture of the papillary muscle would be uncommon causes of sudden cardiac death and would most likely each present with their associated symptoms.

(B) Embolus to the right middle cerebral artery would produce a stroke and not sudden cardiac death.
A 68 year old man with chronic hypertension dies from a myocardial infarction. An autopsy is done and the pathologist finds the a heart similar to the one depicted above. What type of hypertrophy is most likely present in this gross specimen?

a. Asymmetric hypertrophy
b. Concentric hypertrophy
c. Eccentric hypertrophy
d. Hypertrophy in series
e. Symmetric hypertrophy

**Correct answer: B**

When there is pressure overload in the case of chronic hypertension, there are new sarcomeres added in parallel. This leads to concentric hypertrophy (choice B) of the myocardium. Hypertension resulting in myocardial hypertrophy increases a patient’s risk for sudden cardiac death secondary to myocardial infarction.

Eccentric hypertrophy (choice C) can also be described as hypertrophy in series (choice D) as sarcomeres are added in series. This results from volume overload and can be seen in dilated cardiomyopathy.

Asymmetric hypertrophy (choice A) is most often seen in hypertrophic cardiomyopathy as it mainly involves the intraventricular septum.

Symmetric hypertrophy (choice E) is not a term used to describe muscle hypertrophy.
A 14-year-old female presents with polydipsia and polyuria. A finger stick test reveals that her glucose level is 220mg/dl. You suspect that her disease has an autoimmune basis. What is the autoimmune target?

- a. Adrenal beta cells
- b. Pancreatic alpha cells
- c. Pancreatic beta cells
- d. Pancreatic delta cells
- e. Pancreatic gamma cells

**Correct answer: C**

Pancreatic beta cells produce insulin, which regulates the glucose level. In diabetes mellitus Type I, these cells are attacked by the body's own antibodies, resulting in a depletion of insulin. Pancreatic alpha cells produce glucagon. Pancreatic delta cells produce somatostatin. Pancreatic gamma cells and adrenal beta cells are not known to exist within the human body.
Lab results from a 30-year-old male show low urinary cortisol, low serum cortisol and no electrolyte abnormalities. Serum ACTH levels are also low. The man presented to his physician 3 days earlier with fatigue and depression. Which of the following is the most likely diagnosis?

a. Cushing's Syndrome  
b. Primary Adrenal Insufficiency  
c. Prolactinoma  
d. Secondary Adrenal Insufficiency  
e. Sheehan's Syndrome

**correct answer: D**

Low urinary and serum cortisol should immediately make you think of adrenal insufficiency. To discriminate between primary and secondary, it is important to evaluate blood ACTH levels. While ACTH levels are high in the Primary type (indicating a failure of the adrenal glands to produce cortisol when stimulated by ACTH), the Secondary type manifests with low ACTH levels (indicating failure of the pituitary gland to produce enough ACTH).
A 31-year-old man presents to the emergency room complaining of weakness, palpitations, diaphoresis, and irritability. He has had several similar episodes in the past few weeks, none as severe as this one. Physical exam is unremarkable. The nurse performs a fingerstick to check his blood glucose, which is found to be 39. Administration of glucose relieves his symptoms. Serum calcium is 11.1. A mass in which of the following structures is likely responsible for his presenting symptoms?

- a. Liver
- b. Pancreas
- c. Prostate
- d. Small intestine
- e. Stomach

**correct answer:** B

This patient's presenting symptoms and hypoglycemia raise the possibility of an insulinoma, although there are other possibilities (adrenal insufficiency, hypopituitarism, hepatic insufficiency). An insulinoma is a tumor of the beta cells of the pancreatic islets. Over 99% of insulinomas originate in the pancreas. They are associated with MEN-I syndrome (pituitary, pancreas, parathyroid neoplasms), which is important in this case because this patient is hypercalcemic, which may be due to parathyroid hyperplasia. Approximately 90% of patients with MEN-I syndrome have parathyroid hyperplasia.

For this patient, C-peptide and proinsulin levels should be checked prior to further studies to rule out the possibility of insulin self-injection. CT and other localizing tests could then be performed. About 80% of insulinomas are benign, solitary adenomas that are cured with surgical resection.
A 10-year-old boy is severely burned. Nitrogen loss occurs during the first few days after the burn. During this acute period, which of the following substances plays a major role in nitrogen loss?

- a. Cortisol
- b. Erythropoietin
- c. Insulin
- d. Parathyroid hormone
- e. Thyroxine (T4)

**Correct answer:** A

When the body is stressed, physiologically or pathologically, cortisol (option A) is produced in greater quantities than normal. In this case, severe burns is the stressor which would lead to an increase in cortisol. As a glucocorticoid, cortisol will increase protein catabolism which will increase nitrogen loss.

Erythropoietin (option B) is released by the kidneys to stimulate RBC production.

Insulin (option C) is released by the pancreas and regulates glucose levels and fat metabolism.

Parathyroid hormone (option D) is released by the parathyroid gland and regulates calcium, phosphate and Vit D.

Thyroxine (option E) released by the thyroid plays a part in increasing metabolic rate but would not specifically be involved in protein catabolism and thus nitrogen loss.
A 66-year-old man has become increasingly short-tempered with his wife. He has diarrhea, weight loss, and weakness in the proximal muscles. He has atrial fibrillation and tachycardia. Which of the following is the most likely diagnosis?

a. Congestive heart failure  
b. Cushing syndrome  
c. Hyperthyroidism  
d. Mitral valve prolapse  
e. Pheochromocytoma

**Correct answer:** C

Hyperthyroidism (choice C) is often due to excessive production of thyroid hormone in the absence of TSH stimulation. Classic symptoms include irritability, nervousness, weight loss, increased appetite, tremor, sweating, and tachycardia.

Congestive heart failure (choice A) is a condition where decreased cardiac output causes inadequate perfusion of tissues. It does not cause irritability.

Cushing syndrome (choice B) is any condition that causes excess serum cortisol. Signs and symptoms include weight gain, round face, thinning of skin, purple striae, and hirsutism.

Mitral valve prolapse (choice D) is a condition where the mitral valve bulges into the left atrium during systole. It would not cause this man's clinical presentation.

Pheochromocytoma (choice E) is a tumor of the adrenal medulla that causes excess secretion of catecholamines, such as epinephrine and norepinephrine.
A 16-month-old African-American infant living in Seattle is seen for a well-child visit. His weight and height are in the 5th percentile for his age. Nutritional history reveals that he was exclusively breast-fed until 7 months of age when iron-fortified cereal was introduced into his diet. His mother admits that she has a difficult time getting him to try new foods. Laboratory values show serum calcium of 7.5 mg/dL, serum phosphorous of 3.0 mg/dL, serum alkaline phosphatase of 300 U/L, and serum 25-hydroxycholecalciferol of 15.0 nmol/L. Physical examination of this patient with risk factors for vitamin D deficiency is unlikely to include which of the following?

a. Delayed walking  
b. Genu valgum  
c. Large, open anterior fontanelle  
d. Palpable enlargement of the costochondral junctions  
e. Springiness to pressure on the skull

**Correct answer:** B

Infants who are primarily breast-fed, dark-skinned, and have rare exposure to sunlight are at risk for vitamin D deficient-rickets. Rickets can become clinically evident in late infancy with the delayed closure of an abnormally large fontanelle (Choice C) and craniotabes (Choice E), which manifests as a “Ping-pong ball” springiness when pressure is applied to the skull due to thinning of the outer layer. Infants with rickets are more likely to have delayed walking and genu varus (bow-legs) rather than genu valgum (knock-knees). Laboratory values may show normal to low serum calcium, low serum phosphorous, elevated serum alkaline phosphatase, and low serum 25-hydroxycholecalciferol. Supplementation is required in children at risk for vitamin D deficiency.
A 12-year-old girl has recently been diagnosed with type I diabetes and is interested in finding out more about the disease. As her physician, you tell her that the hormone she is deficient in is made by which of the following cell types?

- a. Epsilon cells
- b. Islet alpha cells
- c. Islet beta cells
- d. Islet delta cells
- e. Pancreatic polypeptide cells

**Correct answer:** C

The pancreas is composed of islets of endocrine cells dispersed throughout the exocrine pancreas. There are four different types of islet cells that produce four different types of hormones. Islet alpha cells (Choice B) produce glucagon, a hormone that increases blood glucose concentrations by increasing gluconeogenesis and glycogenolysis in the liver. Islet beta cells (Choice C) produce insulin, a hormone that moves glucose into insulin-sensitive tissues and promotes its storage as glycogen by increasing the rate of glycogen synthesis and decreasing the rate of glycogenolysis. In type I diabetes, autoimmune destruction of the beta cells results in insulin deficiency. Overt diabetes does not appear until approximately 90% of beta cells are destroyed. Islet delta cells (Choice D) produce somatostatin, a hormone that inhibits the secretion and reduces the effects of both insulin and glucagon. Epsilon cells (Choice A) produce ghrelin, a hormone that stimulates appetite. Pancreatic polypeptide cells (Choice E) produce pancreatic polypeptide, a hormone whose function is largely unknown, but is thought to suppress pancreatic secretion and stimulate gastric secretion.
A 2 year old child is brought to the pediatrician for failure to thrive. The child is short, with coarse facial features, and a protruding tongue. The child has not met appropriate developmental milestones. Deficiency of which of the following is most likely to explain these findings?

a. Cortisol
b. Norepinephrine
c. Somatostatin
d. Thyroxine (T4)
e. Insulin

**Correct answer:** D
Choice D is correct

This child has cretinism, a condition that is uncommon in the U.S. given routine testing and treatment at birth. Without treatment, congenital lack of thyroid hormones will lead to severely stunted physical and mental growth.

Choice A is incorrect
A lack of cortisol from primary adrenal failure leads to Addison disease.

Choice B and C are incorrect
There is no deficiency caused by a lack of these compounds

Choice E is incorrect
A lack of insulin leads to Type I diabetes mellitus
A 20 year old female presents to clinic complaining of "whitish" breast secretions over the past month. She is concerned because her last menstrual period was 3 months ago, though she denies any previous or current sexual activity. A urine B-HCG test run in the office is negative. Physical examination is significant only for scant white discharge coming from both breasts. Brain imaging reveals a 0.6cm mass near the pituitary gland. Which of the following is most likely to result from this patient’s condition?

- a. Hyperthyroidism
- b. Cushing disease
- c. Infertility
- d. Acromegaly
- e. SIADH

**correct answer:** C

This stem describes a classic presentation of a prolactinoma- the most common hormone secreting pituitary tumor. Prolactinomas lead to galactorrhea, amenorrhea and infertility secondary to excessive prolactin secretion which (through a feedback loop) inhibits secretion of other gonadotropins causing hypoestrogenism. Acromegaly (choice D) can result from a growth hormone secreting pituitary tumor after fusion of the growth plate in adults. Cushings disease (choice B) would result from an ACTH secreting pituitary tumor, and would present with the classic buffalo hump, central obesity, hirsutism, etc, different from the symptoms seen in this patient. Syndrome of Inappropriate ADH secretion- SIADH (choice E) is not usually caused by a pituitary tumor, but is often a paraneoplastic syndrome or sequelae of brain injury or infection. TSH producing pituitary adenomas leading to hyperthyroidism (choice A) are rare and would cause symptoms different than those seen in this patient including heat intolerance, weight loss, tremors, feelings of anxiety, etc.
A 48-year-old woman from Guatemala complains of epigastric pain. Which of the following clinical settings is most consistent with a duodenal ulcer?

- a. Chronic NSAID use
- b. Crohn's disease
- c. Gastric carcinoma
- d. H. pylori infection
- e. Pernicious anemia

correct answer: D

H. pylori is implicated in almost all cases of duodenal ulcers. Its incidence is very high in developing countries. Chronic NSAID use and increased pain after meals are associated with gastric ulcers. Crohn's disease has no association with peptic ulcer disease. Gastric carcinoma is considerably less likely to result in ulceration. Pernicious anemia, autoimmune destruction of the gastric parietal cells, results in atrophic gastritis and would lead to decreased acid secretion.
An otherwise healthy 3-week-old boy is brought to the physician's office because of jaundice and dark urine for the past 2 weeks. He has hepatomegaly, and his stools are loose, clay-colored, and acholic. Serum conjugated bilirubin concentration is increased. Which of the following is the most likely cause of the hyperbilirubinemia?

a. Defect in cholesterol synthesis
b. Deficiency of glucuronosyltransferase
c. Hemolysis
d. Inflammation of the terminal ileum
e. Obstruction of the biliary system

correct answer: E

The key point here is the increase in conjugated bilirubin, demonstrating that the liver is functioning properly. Combined with hepatomegaly, jaundice, bilirubinuria, and the described abnormal stools, this conjugated hyperbilirubinemia suggests an obstruction of the biliary system (E).

A defect in cholesterol synthesis (A) could adversely affect bile acid production and result in acholic, loose, clay-colored stools. However, such a defect should not affect bilirubin excretion and thus would not explain the hyperbilirubinemia seen in this infant. Furthermore, these defects in cholesterol synthesis are much less common than obstructions of the biliary system (E).

A deficiency of glucuronosyltransferase (B) is suggestive of Gilbert's syndrome (GS) or the much less common Crigler-Najjar syndrome (CDS). However, in GS and CDS the hyperbilirubinemia is due to unconjugated, not conjugated, bilirubin. Unconjugated bilirubin is insoluble, and therefore would not appear in the urine.

Hemolysis (C), like the glucuronosyltransferase disorders, results in an unconjugated, not conjugated, hyperbilirubinemia. Again, unconjugated bilirubin would not be seen in the urine.

Inflammation of the terminal ileum (D) in an infant with loose, clay-colored stools suggests celiac disease. Though iron absorption (and many other substances) may be affected, celiac disease should not cause hyperbilirubinemia. Crohn's disease would also affect the terminal ileum and may have symptoms of malabsorption, but it too should not cause hyperbilirubinemia. Further, onset of Crohn's is usually not seen until 15 – 30 years of age.
A 30-year-old woman has anxiety about episodes of abdominal pain that have alternated with diarrhea and constipation over the past year. She often has these episodes when she is stressed or tired. Physical examination and laboratory studies are within normal limits during these episodes. Which of the following is the most likely diagnosis?

a. Gastroenteritis
b. Generalized anxiety disorder
c. Hypochondriasis
d. Irritable bowel syndrome
e. Major depressive disorder

**Correct answer:** D

(D) The most likely answer is irritable bowel syndrome. Alternating periods of constipation/diarrhea associated with psychosocial stress is typical of IBS. Physical and laboratory studies are normal in IBS.

(A) Gastroenteritis generally follows more of an acute timeline, does not wax and wane, constipation is not typically a component of the illness, and gastroenteritis implies that there may be nausea/vomiting from stomach involvement.

(B) Generalized anxiety disorder involves chronic anxiety often with little to provoke the anxiety and may include physical symptoms, but alternating periods of diarrhea/constipation is not a common complaint. Additionally, patients with GAD will tend to complain of anxiety related to a variety of everyday problems. This woman's anxiety is related specifically to her GI complaint.

(C) We are not given any information to lead us to a diagnosis of hypochondriasis. She has no history of multiple visits to the doctor or hospital with undiagnosed problems.

(E) We are not given information that would lead us to a diagnosis of major depressive disorder. This woman's anxiety is related to her GI complaints.
A 58 year old woman with a long history of alcoholism presents with diarrhea, general weakness, and a 9lb weight loss in the past 3 months. Further history reveals that she has been having difficulties driving at night as well as episodic upper abdominal pain not relieved with antacids or proton pump inhibitors. Laboratory studies reveal a serum ionized calcium of 3.5mg/dL (N=4.5-5.6mg/dL); PT of 30 seconds (N=11-15seconds); PTT of 60 seconds (N=25-40seconds). What is the most likely etiology of this patient’s diarrhea?

a. Carcinoid tumor
b. Pancreatic cancer of the head of the pancreas
c. Chronic pancreatitis
d. Chronic gastritis
e. Celiac sprue

**Correct answer:** C

The patient’s clinical picture most likely reflects a malabsorption syndrome. Chronic pancreatitis (choice C) resulting in chronic pancreatic insufficiency is the most likely diagnosis. Chronic inflammation of the pancreas can result in a critical amount of damage to both the endocrine and exocrine functions of the pancreas. Without the exocrine function of the pancreas, dietary lipids cannot be digested and absorbed nor can the fat-soluble vitamins D, A, K, E. Malabsorption of lipids will result in an osmotic steatorrhea. Malabsorption of vitamins D, A, K, E will result in their respective deficiency symptoms (D=hypocalcemia; A=night blindness; K=prolonged PT/aPTT)

Carcinoid tumor (choice A) can cause carcinoid syndrome which is characterized by secretory diarrhea, flushing, and right-sided heart failure.

Pancreatic cancer (choice B) involving the head of the pancreas can produce a similar clinical picture; however, one would also expect to see signs of obstructive jaundice and elevated LFTs.

Chronic gastritis (choice D) is common in alcoholic patients and can explain the epigastric pain; however, it does not commonly cause malabsorption.

Celiac sprue (choice E) is a common cause of malabsorption. The condition is associated with hypersensitivity to dietary gluten; however, in the setting of an alcoholic, pancreatic insufficiency secondary to chronic pancreatitis is a more likely diagnosis.
A 55-year-old male is complaining of increasing right upper quadrant pain and jaundice. His physical exam is notable for mild scleral icterus, a 12 cm liver span and vague right upper quadrant pain on palpation. His laboratory values are as follows:

- AST: 454
- ALT: 206
- Alkaline phosphatase: 126
- Gamma GT: 174
- Total bilirubin: 2.5
- Direct bilirubin: 1.8

What is the most likely diagnosis?

a. Acetaminophen overdose
b. Alcoholic steatohepatitis
c. Ascending cholangitis
d. Non-alcoholic steatohepatitis (NASH)
e. Viral hepatitis

**Correct answer:** B

In alcoholic steatohepatitis (option B), the most notable feature is that the AST:ALT>2 and that the AST is usually about 500. In addition the patient will sometimes present as jaundiced with RUQ tenderness. In the beginning stages of cirrhosis the liver is enlarged, but as the liver becomes more scarred and fibrotic, the liver span is often difficult to estimate. In viral hepatitis (option E) the AST and ALT rise about the same and the levels are often above 500. In comparison to acetaminophen overdose (option A) where the liver enzymes will be increased significantly, often >1000.

NASH (option D) is usually a diagnosis of exclusion. It presents in a similar fashion of alcoholic steatohepatitis, but there is no history of alcohol abuse, the AST:ALT<2 and the GGT is usually not greater than the alkaline phosphatase.

Ascending cholangitis is recognized by Charcot's Triad: fever, jaundice and right upper quadrant pain. In addition the AST and ALT can be increased, but usually they are not to levels seen in this patient and there is no ratio of 2:1 seen.
A 50-year-old female is admitted for fever of 39 and "8 out of 10" abdominal pain. She complains of diarrhea that she describes as light colored stools that are foul smelling. On physical examination, she is noted to be jaundiced and have tenderness in her right upper quadrant of her abdomen. Labs are sent and are as follows:

- **WBC**: 15,000 with 10% bands
- **Hgb**: 13.3
- **Platelets**: 230,000
- **Alk phos**: 300
- **Total bilirubin**: 7.4
- **Direct bilirubin**: 6.3

What is the most likely cause of this patient's condition?

- a. Ascending cholangitis
- b. Cholecystitis
- c. Choledocholithiasis
- d. Gastroenteritis
- e. Hereditary spherocytosis

**Correct answer**: A

The patient shows the classic signs of ascending cholangitis (fever, jaundice and right upper quadrant pain) called Charcot’s Triad. Ascending cholangitis is caused by chronic obstruction, usually by gallstones (choledocholithiasis, option C) that becomes infected. Fever and increased WBC counts are signs of infection that distinguish cholangitis from choledocholithiasis. Diarrhea of light colored stools that are foul smelling is steatorrhea (fatty stool) since bile is unable to enter the GI tract and digest fat.

Cholecystitis (option B) is an infection of the gallbladder which will present with fever and right upper quadrant pain (Murphy's sign), but usually will not present with jaundice since bile flow is not obstructed. Although the gallbladder is obstructed due to a stone in the cystic duct, the common bile duct is clear and bile can flow directly from the liver to the GI tract.

Gastroenteritis (option D) will present with fever and diarrhea, but will usually present with vomiting, and there is rarely any jaundice or rise in bilirubin.

Hereditary spherocytosis (option E) is a autosomal dominant condition of the membrane of RBCs that causes hemolytic anemia. In hemolytic anemia, the lysis of RBCs will cause an increase in indirect bilirubin due the breakdown of hemoglobin. In this case, the jaundice is caused by direct bilirubin and therefore cannot be attributed to a hemolytic anemia.
A newborn child is brought to the attention of the pediatrician since he has still failed to pass meconium in the first 72 hours and that there is no stool in the diaper. On examination, the infant has a notably distended abdomen, and there is good anal tone. A pilocarpine sweat test is within normal limits. An ultrasound of abdomen is within normal limits. A barium enema shows a large dilated segment of transverse colon.

What is most likely cause for this infant's constipation?

a. Defective CFTR transporter  
b. Duodenal atresia  
c. Lack of ganglion cells in distal colon  
d. Telescoping of bowel segment  
e. Hypertrophic pylorus

**Correct answer: C**

This infant is suffering from Hirschsprung's disease, which is congenital megacolon caused by a lack of Auerbach's and Meissner's plexuses of the distal colon (option C). Infants with Hirschsprung's often will not pass meconium in the first couple of days in the hospital and will continue to have constipation. A barium enema showing a dilated colon followed by a biopsy showing a lack of ganglion cells is diagnostic of the disease. The only treatment is surgical removal of the affected colon.

Although the child did not pass meconium (possible meconium ileus), he is not suffering from cystic fibrosis (defective CFTR transporter, option A) since his sweat test is within normal limits.

Duodenal atresia (option B) and hypertrophic pylorus (pyloric stenosis, option E) would present with vomiting with feeding rather than constipation.

Telescoping of bowel segment (option D) is the cause of intussusception. Intussusception would cause a bowel obstruction and therefore constipation. Any passed stool is likely to be bloody, classically described as "current jelly." However, this condition would not appear with failure to pass meconium (most commonly presenting between five months and three years) and a barium enema would show the overlapping segment of bowel and is the usually the first line treatment (along with air-contrast enema).
A 3-year-old girl from Finland is at the 10th percentile on the growth curve for height and weight. Her parents report that she frequently has foul-smelling diarrhea, particularly after eating pasta or bread. Physical exam reveals excoriated papules and vesicles on her elbows, knees, back, and buttocks. Her endoscopic small bowel biopsy is shown above. Given her condition, for which gastrointestinal neoplasm is she likely at an increased risk?

a. Adenocarcinoma
b. Carcinoid
c. Gastrointestinal stromal tumor
d. Hamartoma
e. T-cell lymphoma

correct answer: E

This patient has the signs and symptoms of celiac sprue, which is an autoimmune-mediated intolerance of gliadin, leading to steatorrhea. Celiac sprue is more common in people of northern European descent. The patient's skin findings are typical of dermatitis herpetiformis, which is often associated with Celiac. Her biopsy shows blunting of the villi, crypt hyperplasia, and lymphocytic infiltration. Celiac patients have a 10-15% chance of getting small bowel T-cell lymphoma (choice E), which is a non-Hodgkin lymphoma that affects T cells.

Celiac patients are also predisposed to adenocarcinoma of the small bowel (choice A), but the risk of T-cell lymphoma is higher.

The other answer choices are GI neoplasms that are not associated with celiac sprue. Carcinoid (choice B) is a serotonin-producing neuroendocrine tumor that can cause carcinoid syndrome.

Gastrointestinal stromal tumor (choice C) is a sarcoma of interstitial cells of Cajal, which are normally involved in controlling gut motility.

Hamartoma (choice D) is a disorganized mass of normal tissue that resembles a neoplasm.
Sickle-cell anemia can be caused by a single amino-acid substitution in the Beta-chain of the hemoglobin molecule. Individuals who are homozygous for this trait will be at greater risk of anemic complications. Infection with which of the following pathogens places the individual at greater risk for an aplastic crisis, specifically?

a. Chlamydia trachomatis
b. Epstein barr virus
c. Parvovirus B19
d. Staphylococcus aureus
e. Trypanosoma cruzi

**Correct answer:** C

The Parvovirus B19 family is known to induce aplastic crisis in individuals that are homozygous for the above HbS/- trait. This virus is known as an 'Erythroivirus' because of its ability to invade red blood cell precursors in bone marrow. NOTE: The same virus is a culprit of the childhood disease, Erythema Infectiosum, and in pregnant women: hydrops fetalis.
An 88-year-old female presents to the ER with generalized jaundice and dark urine. A blood sample is taken and reveals dramatically increased levels of conjugated bilirubin. What is the likely diagnosis?

- Billiary obstruction
- Glucuronyl transferase deficiency
- Hematoma
- Increased hemolysis
- Reduced hepatic bilirubin uptake

**Correct answer:** A

The increase of conjugated bilirubin rules out the causes of unconjugated hyperbilirubinemias (glucuronyl transferase deficiency, hematoma, and increased hemolysis). Reduced hepatic bilirubin uptake would result in decreased blood levels, not increased. Crigler-Najjar syndrome and Gilbert syndrome are examples of glucuronyl transferase deficiencies.
A 30-year-old woman is admitted to the hospital with progressive fatigue and weakness with dyspnea on exertion. On physical examination the following is found: temperature of 39 deg Celsius, pulse: 110 bpm, respiratory rate: 16/min, and blood pressure: 110/70 mmHg. The woman has profound scleral icterus and lymphadenopathy. Blood count reveals: WBC: 9,000/ul, hemoglobin: 5.3 g/dl, Hct: 16%, RBC: 1,600,000/ul, platelet count: 300,000/ul, reticulocyte count: 15%. Which of the following types of anemia is the most likely cause for the patient's presentation?

- a. Anemia of Chronic Disease
- b. Aplastic Anemia
- c. Hemolytic Anemia
- d. Megaloblastic Anemia
- e. Sickle Cell Anemia

**Correct answer:** C

You should immediately try to identify abnormal lab values to develop a sense for what is happening in this patient. The most striking abnormalities are in the low Hgb count, low Hct, and low levels of RBCs in the bloodstream. Such a scenario indicates either 1) increased destruction of RBCs or 2) decreased production of RBCs. To evaluate production of RBCs, it is useful to look at the reticulocyte count. An elevated reticulocyte count (here, 15%) means that the bone marrow is responding appropriately to the anemia, suggesting that the low RBC count is due to blood cell destruction rather than insufficient production. Further, hemolytic anemia will often result in the release of unconjugated bilirubin (from Hgb) into the bloodstream. In our patient, this presents itself as scleral icterus.
Hospital discharge of a 75-year-old man is delayed due to unavailability of a bed in a nursing home. He is bedridden and unable to attend to his personal needs. During a 3-day period, his pulse increases from 82/min to 125/min, and blood pressure decreases from 124/72 mm Hg to 100/55 mm Hg. Laboratory values are above. Which of the following is the most likely diagnosis?

- a. Acute renal failure
- b. Dehydration
- c. Diabetic ketoacidosis
- d. Gastrointestinal hemorrhage
- e. Syndrome of inappropriate ADH (vasopressin)

**Correct answer:** B

The increased pulse and decreased blood pressure indicate a loss of fluid (volume contraction). The patient's hemoglobin, urea nitrogen, and sodium all rise to above the normal values by Day 3, whereas his glucose and creatinine remain normal. These changes are consistent with an hyperosmotic volume contraction, a loss of water without losing solute. This typically occurs due to dehydration.

Choice A is incorrect because renal failure would decrease creatinine clearance.

Choice C is incorrect because diabetic ketoacidosis would cause increased blood glucose levels.

Choice D is incorrect because gastrointestinal hemorrhage would present with additional symptoms and hemoglobin would decrease.

Choice E is incorrect because SIADH would cause decrease in urine volume and retention of fluid, not loss of fluid.
A 68-year-old man has had low back pain over the past 2 months. Laboratory studies show a normochromic, normocytic anemia and azotemia. Serum and urine calcium concentrations are abnormally increased, and urinalysis shows excessive protein (4+) and proteinaceous casts. Bone marrow examination is most likely to show uncontrolled proliferation of which of the following cells?

a. Basophils  
b. Lymphocytes  
c. Macrophages  
d. Plasma cells  
e. Reticulocytes

**Correct answer:** D

The patient described has multiple myeloma, which is characterized by proliferation of plasma cells and hyperproduction of monoclonal light chains (Bence-Jones proteins). Accumulation at the renal tubules causes renal insufficiency. MM also causes bone destruction, both from marrow expansion and stimulation of osteoclasts. This causes hypercalcemia, which also contributes to renal dysfunction. As with any leukemia, other cell lines are suppressed, causing anemia and thrombocytopenia.
A 75-year-old woman has increasing shortness of breath on exertion. Findings on physical examination are unremarkable. X-rays of the chest show no abnormalities of the heart or lungs. Pertinent laboratory findings include Hematocrit 28%, Hemoglobin 9 g/dL, Mean corpuscular volume 70 micrometer$^3$

Which of the following is the most likely basis for these findings?

a. Acquired hemolytic anemia  
b. Chronic blood loss  
c. Folic acid deficiency  
d. Beta-Thalassemia minor  
e. Pernicious anemia

**correct answer:** B

The lab values for hematocrit and hemoglobin indicate that this women has anemia. Her MCV value of 70 is consistent with a microcytic anemia. The most common microcytic anemia is iron deficiency anemia. Chronic blood loss through the GI tract is the most common cause of iron deficiency anemia in men and post menopausal women.

An acquired hemolytic anemia likely would present with additional symptoms on physical exam, including signs of jaundice, dark urine upon standing, and splenomegaly. Also, the history does not suggest a cause for hemolysis.

Beta-Thalassemia minor is a mild microcytic anemia. In the majority of cases, Beta-Thalassemia minor is asymptomatic, and many affected people are unaware of the disorder. This is an inherited anemia that would not present late in life with increasing shortness of breath.

Folic acid deficiency and Pernicious anemia leading to Vitamin B12 deficiency are both types of macrocytic and megaloblastic anemia. The MCV value would be elevated above 96 micrometer$^3$ and these types of anemia are less common than iron deficiency anemia.
A 47 year old woman presents to the ED complaining of a swollen left leg. The swelling began two days ago and occasionally causes pain. The patient has a history of breast cancer that is currently being treated with doxorubicin and paclitaxel. She recalls no significant family history. On exam the left leg is edematous below the knee with mild erythema; the right leg is normal. A venous cord is palpated just superior to the medial malleolus. Upon dorsiflexion of the left ankle, the patient winces in pain. Which of the following is a risk factor for the most likely cause of this patient’s symptoms?

   a. Female gender  
   b. Paclitaxel  
   c. Breast cancer  
   d. Knee arthritis  
   e. Age of 47

**Correct answer:** C

This patient likely has a deep vein thrombosis (DVT). Whenever you see DVT, think Virchow's triad. Virchow's triad describes the most common causes of thrombosis - endothelial injury, stasis, and thrombophilia.

Cancer is a proinflammatory state that strongly predisposes towards thrombosis, and is an established major risk factor for DVT.

Female gender is not by itself a risk factor for DVT. However, estrogen therapy (whether OCPs or estrogen replacement) can be a risk factor in women.

Paclitaxel is an antineoplastic drug that targets microtubules, but does not predispose to thrombosis.

Knee arthritis does not predispose to DVT, unless it was so incapacitating as to lead to immobility.

Age of 47 is not a risk factor. Older age (>60) is a minor risk factor for DVT. Any unprovoked DVT in a younger person is an indication for a thrombosis workup.
A 29-year-old woman visits an ophthalmologist after noting a gradual decrease in visual acuity in her right eye over the last year. She plays tennis 3 times per week, eats a low-fat diet, and has a BMI of 24.4. She denies headache, trauma to the eye, or a family history of diabetes or multiple sclerosis, and does not take any medications. She is concerned because her father and paternal grandmother suffered from early blindness. On exam, the ophthalmologist notes several exudative hemangiomas on the retina of her right eye. A CT scan of her head reveals bilateral cerebellar cysts and medullary hemangioblastomas. This patient is at high risk of developing which of the following?

a. Angiomyolipoma
b. Cystic fibrosis
c. Renal cell carcinoma
d. Retinitis pigmentosa
e. Scleroderma

correct answer: C

This patient has Von Hippel-Lindau disease (VHL), caused by an autosomally dominant germline mutation to the VHL tumor suppressor on chromosome 3. Visual changes due to retinal damage are often the presenting symptom, and occur most frequently in the third decade of life. Up to 50% of patients with Von Hippel-Landau syndrome go on to develop renal cell carcinoma. Pheochromocytoma, pancreatic tumors, and papillary cystadenomas of the broad ligament are also seen with some frequency.

None of the other answer choices is associated with Von Hippel Lindau syndrome.
An 8-year-old male presents at a clinic in a rural Nepal with conjunctival scarring and ocular pruritus. His physicians are concerned with preventing progression to blindness.

Which of the following is the most likely underlying etiology?

a. Cataracts
b. Chlamydia trachomatis infection
c. Congenital malformation
d. Macular degeneration
e. Trauma

**Correct answer:** B

Infection with Chlamydia trachomatis is the leading cause of preventable blindness in the world.

It is unlikely that a congenital malformation would result in conjunctival scarring and ocular pruritus.

Cataracts, macular degeneration or trauma are not suggested by the patient’s medical history, and should not be suspected without more information.
An 18-year-old female athlete reports easy fatigability and weakness. Physical examination shows no abnormalities. Laboratory studies are above. Which of the following is the most likely diagnosis?

a. Aldosterone deficiency  
b. Anxiety reaction with hyperventilation  
c. Diabetic ketoacidosis  
d. Ingestion of anabolic steroids  
e. Surreptitious use of diuretics

**Correct answer:** E  
This patient’s serum bicarbonate is high, while her potassium is low. The potassium level in her urine is high. (Typical range is 50-150mEq per day). This indicates the use of a diuretic which blocks the reuptake of potassium.

Choice A is incorrect because aldosterone increases potassium secretion, therefore aldosterone deficiency would cause decreased potassium secretion.

Choice B is incorrect because hyperventilation causes respiratory alkalosis, resulting in decreased bicarbonate in the serum.

Choice C is incorrect because acidosis causes decreased potassium secretion. Also, diabetic ketoacidosis would cause additional symptoms and abnormally high blood glucose levels.

Choice D is incorrect because ingestion of anabolic steroids generally does not cause the type of potassium imbalance indicated by the lab values.
A 74-year-old man has had confusion for 2 weeks. He has smoked two packs of cigarettes daily for 50 years. An x-ray of the chest shows a 5-cm mass in the lung. Laboratory studies of serum are above. Which of the following is the most likely cause of these findings?

a. Adenocarcinoma of the lung
b. Craniopharyngioma
c. Medullary carcinoma of the thyroid gland
d. Renal cell carcinoma
e. Small cell carcinoma of the lung

**Correct answer:** E

The first thing you need to recognize to answer this question is that this patient has hyponatremia (110 mEq/L, normal is between 135-145). Given the clinical finding of a lung mass, the most likely explanation is that this patient has SIADH, a paraneoplastic complication of small cell carcinoma of the lung (choice E). SIADH is syndrome of inappropriate antidiuretic hormone (ADH) secretion, in which the body, or in this case the lung tumor, secretes ADH that is not responsive to normal feedback mechanisms, resulting in inappropriate retention of water and dilution of the sodium concentration.

The other malignancies in the answer choices are not associated with SIADH. Adenocarcinoma of the lung (choice A) is more common in non-smokers and is not associated with SIADH.

Craniopharyngioma (choice B) is usually a tumor of childhood that is a remnant of Rathke’s pouch; it could compress the pituitary but this would lead to decreased ADH production, not increased.

Medullary carcinoma of the thyroid (choice C) could secrete excess calcitonin, but not ADH.

Renal cell carcinoma (choice D) can secrete excess erythropoietin, leading to polycythemia, or excess red blood cells, but this would have no effect on sodium concentration.
A 16-year-old boy has a 1-day history of pain in the right ear. He swims every morning. The right ear canal is red and swollen. He has pain when the auricle is pulled or the tragus is pushed. Which of the following is the most likely diagnosis?

- a. Acute otitis media
- b. Bullous myringitis
- c. Chronic otitis media
- d. External otitis
- e. Mastoiditis

correct answer: D

Ear pain worsened on manipulation, with inflammation of the canal (i.e. external ear) is most consistent with external otitis (D), colloquially known as “swimmer’s ear.”

Acute otitis media (A) is an infection of the middle ear that usually presents with earache unaffected by external manipulation, with a red, bulging tympanic membrane on exam. Patients are often febrile to 102 degrees F (38.9 degrees C) or more. Children under 7 are more susceptible to this condition due to their shorter, more horizontal pharyngotympanic (aka auditory or Eustachian) tubes.

Bullous myringitis (B) is an infection of the tympanic membrane (TM) itself, in which bubbles filled with blood form on the TM surface.

Chronic otitis media (C) is incorrect both because of symptoms inconsistent with otitis media (see above), and the 1-day time course of the complaint is more suggestive of an acute process.

Mastoiditis (E) typically manifests with pain, tenderness, and/or swelling of the mastoid process. Further, such a short history of pain is inconsistent with mastoiditis, as it is usually a sequela of otitis media. Accordingly, children (not adolescents) are most frequently affected.
A 25-year-old woman has a 3-day history of vomiting and diarrhea. She has postural hypotension and poor tissue turgor. Her serum sodium concentration is 130 mEq/L. Which of the following findings is most likely?

   a. Decreased serum aldosterone concentration
   b. Increased serum atrial natriuretic peptide concentration
   c. Increased effective circulating volume
   d. Increased serum ADH (vasopressin) concentration
   e. Urine osmolality less than serum osmolality

**Correct answer: D**

ADH (vasopressin) is secreted by the posterior pituitary gland. Its secretion is in response to either activation of baroreceptors in the veins, atria and carotid bodies in response to reduced plasma volume, or by activation of osmoreceptors in the hypothalamus in response to increased plasma oncotic pressure. The patient in this question has orthostatic hypotension and poor skin turgor, clinical signs suggesting volume depletion. Likewise, plasma volume is low and ADH levels are responsively increased. Because plasma sodium concentration is lower than normal, the renin-angiotensin-aldosterone system will be activated and the concentrations of all three will be increased. ANP is secreted by atrial myocytes in response to increased blood pressure, the opposite of this patient. In dehydrated states, urine osmolality is higher than plasma/serum osmolality due to the action of ADH increasing permeability of the collecting duct to reabsorb water.
A 24-year-old woman who breast-feeds her infant develops a warm, painful mass and erythema of the overlying skin in the upper outer quadrant of the right breast. The pain and redness resolve with antibiotic therapy. Several months later, the same area is firm and the overlying skin is dimpled. Which of the following is the most likely cause of these findings?

a. Breast abscess with scarring
b. Fibroadenoma
c. Fibrocystic change
d. Infiltrating mammary carcinoma
e. Traumatic fat necrosis

correct answer: A

A warm, painful, and erythematous breast mass (often accompanied by fluctuance), experienced by breast-feeding women, that resolves with antibiotics suggests an infectious cause. Although far less common than mastitis, a breast abscess is the only infectious etiology among the answer choices. Risk factors include primiparity, maternal age >30, and a pregnancy lasting longer than 41 weeks. Fibrosis (scarring) can replace the abscess in some women, leaving a firm mass and dimpled overlying skin months after the abscess has resolved.
A 37-year-old African-American woman presents with fever, anorexia, purple plaques on her shins, dyspnea on exertion, anterior uveitis, and an asymmetric smile. Which of the following MUST be seen for diagnosis of her disease?

a. Arthralgias
b. Bilateral hilar adenopathy
c. Cardiac conduction abnormalities
d. Increased serum levels of ACE
e. Non-caseating granulomas

correct answer: E

This woman has symptoms suggesting sarcoidosis. Of the options, only non-caseating granulomas (choice E) are required for the diagnosis of the disease. Sarcoidosis is a chronic systemic granulomatous disease often involving multiple organ systems. It occurs most often in female African-Americans. Features include constitutional symptoms, erythema nodosum, dyspnea on exertion, anterior uveitis, and Bell’s palsy, amongst others.

Arthralgias (choice A) are present in 25-30% of cases. Bilateral hilar adenopathy (choice B) is a hallmark of the disease, but is only seen in 50% of cases. Cardiac involvement (choice C) is seen in 5% of cases. Increased serum levels of ACE (choice D) are seen in 50% of cases.
A 30-year-old female with Crohn's disease complains of increasing fatigue over the past several months. She denies any recent flares, and says that she has not seen any blood in her stool. She eats a well balanced diet and takes a multivitamin regularly. Her surgical history is notable for an ileocolic resection 3 years ago. Physical exam shows no abnormal findings, and her fecal occult blood test is negative. What is the most likely cause of this patient's fatigue?

a. B12 deficiency  
b. Calorie deficiency  
c. Folate deficiency  
d. Iron deficiency  
e. Protein deficiency

**Correct answer: A**

This patient is suffering from Crohn's disease (which commonly affects the terminal ileum) and had her ileum resected three years ago; therefore, the most likely cause of her fatigue is anemia caused by a B12 deficiency (option A). B12 is absorbed in the ileum, and therefore, she can no longer absorb B12 from her diet. The human body usually stores enough B12 for a couple of years, which is why she is presenting three years after surgery. The treatment for this condition would be B12 injections. Those without an ileum can also suffer from diarrhea and steatorrhea (fatty stools) since the ileum is where most of the bile acids are reabsorbed. Bile acids are necessary for fat absorption.

Any possible cause of anemia a patient with Crohn's is iron deficiency (option D) due to flares characterized by inflammation and intestinal bleeding. However, she denies any recent flares and her heme-occult test is negative. In addition, iron is absorbed in the duodenum which has not been removed from this patient.

Calorie (B), protein (E) and folate (C) deficiencies are unlikely because she says that she is able to eat a well balanced diet and take a multivitamin. Although she is missing her ileum, she has enough of her small intestine to absorb the necessary nutrients.
A 22-year-old female with no significant past medical history presents to her primary care physician complaining of a painless lesion she discovered on her vulva the day before. Upon further questioning she admits to unprotected sex with a new partner. Her blood work reveals that her HIV ELISA is negative but her VDRL and FTA results are positive. If this patient were to go untreated, which of the following sequelae would develop within 1-6 months of the primary infection?

a. Arthritis
b. Maculopapular rash
c. Pelvic inflammatory disease
d. Shingles
e. Paresis

_correct answer: B_

The spirochete Treponema pallidum causes the chronic systemic infection of syphilis, which is transmitted primarily through sexual contact. Primary syphilis manifests as chancres – or painless, elevated ulcers – that develop approximately 3 weeks after inoculation and heal within 2 to 6 weeks. If untreated, the secondary stage of syphilis manifests as a maculopapular rash (Choice B) on the palms and soles. After resolution of the second stage, the infection enters a latent phase that can last for years. Tertiary syphilis is characterized by gummas of the skin and bones, cardiovascular syphilis with aortitis, and neurosyphilis with menigovascular disease, paresis (Choice E), and tabes dorsalis. Although unlikely, tertiary syphilis could present within 6 months but only if the patient were immunosuppressed (HIV/AIDS, or on immune suppressive drugs). However, the patient has no past medical history and does not have active HIV, so tertiary syphilis is very unlikely.

Untreated infection with Neisseria gonorrhoeae can lead to arthritis (Choice A).

Pelvic inflammatory disease (Choice C), or acute salpingitis, generally begins as a Neisseria gonorrhoeae or Chlamydia trachomatis cervicitis that ascends the female reproductive tract if untreated.

Shingles (Choice D) is a viral disease characterized by painful blisters in a dermatomal distribution. It is caused by reactivation of a previous varicella zoster virus infection.
A 50 year-old female who is being treated with a multidrug regimen for tuberculous pneumonia presents to a clinic complaining of pain and swelling in both arms and legs for the last two weeks. Physical exam is remarkable for a maculopapular rash across her nose and cheeks, and swollen, tender elbows and knees. Which of the following medications is likely responsible for her symptoms?

a. Ethambutol
b. Isoniazid
c. Pyrazinamide
d. Rifampin
e. Streptomycin

**correct answer:** B

This patient likely has drug-induced systemic lupus erythematosus (SLE). Isoniazid (INH) (choice B) is one of four drugs that classically causes this condition, and it fits this patient’s current drug regimen. The other three are hydralazine, procainamide, and phenytoin. INH may additionally result in peripheral parasthesias, which is usually prevented by supplementing its use with vitamin B6.

Pyrazinamide (choice B), Rifampin (choice D) and Streptomycin (choice E) are anti-TB drugs that may cause elevated liver enzymes. Ethambutol (choice A) is another anti-TB drug that may cause optic neuritis.
A 18-year-old male is mugged and hit on the head with a blunt object. He loses consciousness for approximately 10 minutes, at which time, he wakes up and walks home. He is fully conscious for 20 minutes but then gradually becomes lethargic. Where is the likely site of his associated skull fracture?

- a. Basilar region
- b. Frontal region
- c. Occipital region
- d. Parietal region
- e. Temporal region

**correct answer:** E

An epidural hematoma is almost always due to a rupture of the middle meningeal artery. When the skull is fractured (almost always the temporal bone), the middle meningeal artery is torn and bleeds into the epidural space. These injuries tend to present with a loss of consciousness followed by a lucid interval of a few minutes to a few hours with subsequent rapid neurologic deterioration due to continued hemorrhage and brain compression. Treatment is almost always neurosurgery to evacuate the hematoma.
The circles represent the size of a patient’s pupils without treatment and following treatment with tyramine and with epinephrine. Which of the following is compatible with the findings shown for the left eye?

a. Blockade of alpha-adrenergic receptors  
b. Blockade of beta-adrenergic receptors  
c. Blockade of muscarinic receptors  
d. Inhibition of cholinesterase  
e. Sympathetic denervation

**Correct answer: E**

Recall that the sympathetic nervous system causes pupillary dilation (mydriasis) via α-adrenergics, while the parasympathetic nervous system constricts the pupil (miosis) via muscarinic cholinergics. The diagram in this question shows that the left pupil is smaller than the right pupil without treatment, which points to either increased parasympathetic stimulation or decreased sympathetic stimulation in the left eye. Adding tyramine, which is an indirect sympathomimetic, has no effect on the left eye, which is consistent with sympathetic denervation of the left eye (choice E).

The treatment with epinephrine had a large effect on the left eye, however, which rules out blockade of alpha-adrenergic receptors (choice A), since blocking the α-adrenergic receptors would have blocked any effect of epinephrine.

Blockade of β-adrenergic receptors (choice B) is incorrect since B-receptors are not involved in pupillary constriction and dilation.

Blockade of muscarinic receptors (choice C) is incorrect because the left eye is constricted without treatment, not dilated as it would be with unopposed sympathetic stimulation.

Finally, inhibition of cholinesterase (choice D) is incorrect because it would not have blocked tyramine from dilating the left eye, since it acts on adrenergics, not cholinergics.
A 48-year-old woman has loss of pain and temperature sensation in the left upper and lower extremities. Which of the following labeled areas in the drawing of the medulla shown is the most likely site of the causal lesion?

a. Region A  
b. Region B  
c. Region C  
d. Region D  
e. Region E

**Correct answer:** C

To answer this question, you need to know that pain and temperature sensation are carried in the spinothalamic tract. Therefore, impaired pain and temperature sensation points to a lesion in the contralateral spinothalamic tract (Region C). Specifically, the spinothalamic tract carries information about pain and temperature from the contralateral side of the body from the dorsal root ganglion, crossing at the anterior white commissure and finally synapsing in the VPL thalamic nucleus.

Region A is the hypoglossal nerve (cranial nerve XII), which controls the tongue muscles.

Region B is the trigeminal nerve (cranial nerve V) which carries pain and temperature from the ipsilateral face.

Region D shows damage to the medullary pyramid which controls the skeletal muscles of the contralateral body.

Region E is the medial lemniscus, which controls proprioception and fine touch of the contralateral body.
A 50-year-old man with a history of alcoholism has difficulty with short-term memory. He is unable to recall the date and cannot remember what he ate for breakfast this morning. He thinks the examiner is a long-lost friend and carries on a conversation with the examiner as if they have known each other for years. His long-term memory appears intact. The patient dies shortly thereafter of a myocardial infarct. Autopsy examination of his brain is most likely to disclose an abnormality involving which of the following?

a. Amygdala  
b. Caudate nucleus  
c. Hippocampus  
d. Locus caeruleus  
e. Mammillary bodies

**Correct answer:** E

This man suffered from Korsakoff syndrome. His history of alcoholism predisposed him to a thiamine (B1) deficiency, a vitamin that is crucial to cellular energy production. A deficiency of this vitamin causes Wernicke-Korsakoff syndrome. Wernicke encephalopathy is characterized by confusion, ocular disturbance, and ataxia of gait. Korsakoff syndrome manifests as loss of short-term memory and confabulation. The lesion of both of these disorders is found in the mammillary bodies (choice E) of the thalamus.

The amygdala (choice A) is an almond-shaped nuclear mass in the medial part of the anterior temporal lobe. It is involved in processing memory and emotion.

The caudate nucleus (choice B) is a nucleus located in the basal ganglia involved in the control of voluntary movement.

The hippocampus (choice C) is a part of the limbic system found in the medial temporal lobe. It is involved in short term memory and spatial navigation.

The locus coeruleus (choice D) is a nucleus located in the brain stem. It is involved in the stress response, and secretes norepinephrine.
A 40-year-old man with a 20-year history of alcohol abuse is brought to the hospital by his friends because he was difficult to rouse. He ate a large meal several hours ago. He is emaciated and lethargic. Examination shows severely restricted horizontal eye movements and ataxia of both upper extremities. The most likely cause of these findings is a deficiency of which of the following nutrients?

- a. Folic acid
- b. Vitamin A
- c. Vitamin B1 (thiamine)
- d. Vitamin B6 (pyridoxine)
- e. Vitamin B12 (cobalamin)

**Correct answer:** C

This patient is exhibiting signs of Wernicke's encephalopathy, a condition of thiamine deficiency. The classic triad of Wernicke's is encephalopathy, ophthalmoplegia in the form of lateral gaze nystagmus, and ataxia. Chronic alcoholics and individuals undergoing TPN without B1 supplements are predisposed to B1 deficiency. Wernicke's may be precipitated by carbohydrate heavy meals because thiamine is a cofactor for pyruvate dehydrogenase and alpha-ketoglutarate dehydrogenase, key enzymes in glucose metabolism. Untreated Wernicke's encephalopathy may progress to Korsakoff syndrome, an irreversible condition characterized by psychosis, anterograde and retrograde amnesia, and confabulation.
A 50-year-old man has had gradually progressive weakness of the hands during the past year. Physical examination shows atrophy of the forearm muscles, fasciculations of the muscles of the chest and upper extremities, and hyperreflexia of the lower extremities. A Babinski sign is present bilaterally. Sensation is intact. Which of the following is the most likely diagnosis?

a. Amyotrophic lateral sclerosis  
b. Dementia, Alzheimer type  
c. Guillain-Barre syndrome  
d. Multiple cerebral infarcts  
e. Multiple sclerosis

**Correct answer:** A

This patient is exhibiting signs of upper and lower motor neuron degeneration, suggesting a diagnosis of amyotrophic lateral sclerosis (ALS). Upper motor neuron degenerative signs include hyperreflexia, increased tone, and positive bilateral Babinski signs. Lower motor neuron degenerative signs include muscle atrophy, areflexia, fasciculations, and flaccid paralysis. Since ALS causes degeneration of both upper and motor neurons, this is the likely diagnosis for this patient.

Alzheimer dementia is not known to correlate with motor neuron loss. Guillain-Barre is usually post-viral illness of the GI tract and usually presents with a rapidly ascending, reversible muscle paralysis. Multiple cerebral infarcts would explain the upper motor signs but not the degeneration of lower motor neurons. Multiple sclerosis usually presents in middle-aged women with optic signs or singular neurologic defects that include peripheral sensory losses, not the universal loss of motor neurons described above.
A mother brings in her 9-month-old son complaining of a three day history of poor feeding. She reports that he has not fed well and has not had a bowel movement in this time. She feeds him home-processed vegetables from their garden. His immunizations are up-to-date. On exam, the infant has a decrease in muscle tone, abdominal distention, and labored breathing. What is/are the most likely etiologic agent(s)?

- Autoimmune antibodies targeting the post-synaptic acetylcholine receptor
- Lead ingestion resulting in toxic plasma level
- Neurotoxin preventing the release of acetylcholine from presynaptic nerve terminals
- Point mutation resulting in defective muscular protein
- Virus resulting in necrosis of anterior horn cells

**Correct answer:** C

This boy likely has infant botulism (C). Ingested spores of the bacterium Clostridium botulinum can germinate in an infant’s gut and produce botulinum toxin that prevents acetylcholine release from presynaptic nerve terminals, thus affecting somatic and autonomic neuromuscular junctions alike. This results in flaccid paralysis, poor feeding, and constipation. Severe neurotoxin poisoning can result in respiratory failure. Infants are particularly sensitive to honey because their immune systems are not fully mature, and cannot prevent spore proliferation.

Autoimmune antibodies targeting the post-synaptic acetylcholine receptor (A) are seen in myasthenia gravis, an autoimmune disease characterized by muscle weakness that worsens as the day progresses.

Lead poisoning (B) can result in lethargy, poor feeding, and constipation. However, there is no given history suggestive of heavy metal ingestion. In addition, lead poisoning typically occur secondary to chronic exposure, so the presentation is either sub-acute or chronic (weeks to months rather than days).

A point mutation resulting in defective muscular protein (D), dystrophin, describes Duchenne muscular dystrophy, an X-linked genetic disease resulting in progressive proximal muscle weakness. The acute onset of this infant’s symptoms is not consistent with DMD.

A virus that results in necrosis of the spinal cord’s anterior horn (E) describes poliomyelitis. The virus is spread by the fecal-to-oral route, and severe cases lead to flaccid paralysis. While this infant has acute-onset decreased muscle tone, which could be consistent with poliomyelitis, this is much less likely because: (1) the virus does not typically affect autonomic nerves of the gut (as the findings of constipation suggest), and (2) his immunizations are up-to-date – poliomyelitis is extremely rare in the United States due to immunizations in the first year of life.
A 60 year old patient presents with intact verbal fluency and is able to construct whole, expressive sentences. However, this patient is unable to follow commands or comprehend spoken dialog. He is diagnosed with a receptive aphasia.

His brain MRI most likely shows a lesion in which of the following areas?

a. Amygdala
b. Broca's Area
c. Frontal Lobe
d. Reticular Activating System
e. Wernicke's Area

**Correct answer:** E

Wernicke's area (located in the left hemisphere) supports comprehension of spoken language (E). Patients with damage in this region will exhibit receptive aphasia.

Productive aphasia, the inability to form fluent sentences, results from damage to Broca's area (B).

The amygdala (A) is considered the seat of fear and emotional processing in the brain; damage here results in Kluver-Bucy syndrome.

Damage to the Frontal Lobe (C) results in personality changes and deficits in concentration.

Damage to the Reticular Activating System (in the pons or midbrain) results in a comatose state.
An 88-year-old man presents with rigidity, diminished facial expression, slowness in walking, and a resting tremor. On physical exam, he also has orthostatic hypotension, and an apparent cerebellar ataxia. He does not have significant impairment in memory or cognitive abilities. Which of the following histological findings would most likely be present on postmortem examination of his brain?

a. Amyloid beta plaques in the hippocampus
b. Glial cytoplasmic inclusion bodies in the substantia nigra, cerebellum, and interomediolateral cell column of the spinal cord
c. Lewy bodies isolated to the substantia nigra
d. Lewy bodies isolated to the cortex
e. Neurofibrillary tangles in the entorhinal cortex

**Correct answer:** B

The patient described has multiple system atrophy, a neurodegenerative disease that in addition to features of Parkinsonism (rigidity, diminished facial expression, slowness in walking (bradykinesia), and a "pill-rolling" tremor at rest) also has features of autonomic dysfunction (orthostatic hypotension) and cerebellar ataxia. Answer B lists the characteristic pathological findings found in multiple system atrophy, but is also the only answer choice in which lesions are found in parts of the brain that would produce all the symptoms described:

1. Parkinsonism, due to lesions in the nigrastriatal dopaminergic system.
2. Autonomic dysfunction, due to lesions in the cathecholaminergic nuclei of interomediolateral cell column of the spinal cord
3. Cerebellar ataxia, due to lesions in the cerebellum.

Answer choices A and E are the expected pathological findings in Alzheimer's disease, which would present with dementia, but not the movement problems described. A good understanding of neuroanatomy is helpful in answering this question, even if you haven’t memorized all the diseases and the pathological findings associated with them. Lesions to the areas of the cortex described would produce cognitive dysfunction, which the patient does not have.

Answer D, which is the pathological finding in dementia with Lewy bodies, would also present with cognitive impairment, and thus can be ruled out using the same logic.

Answer C is the classic description of pathological findings in Parkinson's disease. (Note: Parkinson’s disease is not the only disease that presents with "Parkinsonism", which this the constellation of symptoms described above related to damage to the substantia nigra, striatum, or drugs that interfere with dopamine such as haloperidol.) This answer choice does not account for the ataxia and the orthostatic hypotension, and thus is less likely to be correct.

A 40-year-old, diabetic female patient comes to see you at your clinic. She has recently taken a new job working as the personal assistant of a high-level executive. Her job requires a lot of computer work, and she frequently works overtime. She complains that she has noticed tingling in both hands that is worse on the left. When you ask her to describe the exact location, she indicates that the first three digits are more numb than the fourth and fifth digits. Irritation of which nerve is the most likely cause of her symptoms?

a. Axillary nerve
b. Median nerve
c. Musculocutaneous nerve
d. Radial nerve
e. Ulnar nerve

Correct answer: B

This patient's symptoms are a classic presentation for carpal tunnel syndrome, in which the median nerve is compressed within the carpal tunnel. This syndrome is frequently caused by overuse, as in this case, with the patient's excess computer use.

The median nerve innervates the lateral three and a half digits on each hand, which explains the numbness in digits 1-3 in her hands. The ulnar nerve innervates the medial digit and a half of the hand, but this patient reported that her digits 4-5 were not numb.

The radial nerve innervates the back of the hand, but not the muscles within the hand. The other nerves do not innervate the hand.
A 24 year-old man presents for his annual check-up and his blood pressure is found to be 152/78. The measurement is repeated and the same result is obtained. His primary care physician suspects secondary hypertension. In which of the following scenarios would this patient have suppression of the renin-angiotensin system?

a. Aortic coarctation
b. Carcinoid syndrome
c. Primary hyperaldosteronism
d. Unilateral renal artery stenosis
e. Volume depletion

**Correct answer:** C

Although unilateral renal artery stenosis, primary hyperaldosteronism, and aortic coarctation are all causes of secondary hypertension, only in primary hyperaldosteronism (or Conn syndrome) would there be suppression of the renin-angiotensin system. In unilateral renal artery stenosis and aortic coarctation, it would be activated due to relative renal hypoperfusion. Volume depletion would not cause hypertension, and the renin-angiotensin system would be stimulated. In carcinoid syndrome (flushing, hypotension, tachycardia, diarrhea), the renin-angiotensin system would be stimulated to increase vascular tone.
One day after a 10-km race, a previously healthy 42-year-old man has dark urine. Urinalysis results are above. Which of the following is the most likely cause of these findings?

a. Acute glomerulonephritis
b. Hypovolemia
c. Renal infarct
d. Renal vein thrombosis
e. Rhabdomyolysis

correct answer: E

Rhabdomyolysis (Choice E) occurs when muscle fibers break down, often due to stress (such as running a race), releasing myoglobin into the bloodstream, which can then damage the kidneys. The recent history of a race, as well as the urinalysis which is positive for blood on dipstick but negative for RBCs supports this diagnosis.

Acute glomerulonephritis (Choice A) usually results in red blood cells in the urine and is unlikely with no history of infection or other symptoms.

Hypovolemia (Choice B) can lead to acute renal failure by prerenal mechanism but would not cause a positive dipstick for blood.

Renal infarct (Choice C) would present with severe pain and renal failure, as would renal vein thrombosis (Choice D).
A 27-year-old HIV positive man comes for a re-visit after a screening urinalysis reveals strikingly elevated protein levels. The patient is currently reluctant to start antiretroviral therapy, though he is taking prophylactic antibiotics. He has a distant history of injection drug use. On exam, the patient has mild ankle edema and is slightly cachectic, but otherwise appears well. A new urinalysis reveals 3+ proteinuria without any hematuria or red cell casts. A serum albumin is 2.3 g. Which disease process is most likely to explain this patient’s proteinuria?

a. Minimal change disease
b. Focal segmental glomerulosclerosis
c. Membranoproliferative glomerulonephritis
d. Membranous nephropathy
e. IgA nephropathy

Correct answer: B

This patient's clinical description is that of nephrotic syndrome. Nephrotic syndrome is characterized by proteinuria without hematuria; other manifestations may include edema, thrombosis, infection, low serum albumin, coagulopathy, and hyponatremia.

Patients with HIV and no other diseases are particularly likely to have a variant of focal segmental glomerulosclerosis (FSGS) called HIV-associated nephropathy (HIVAN). This is especially true among black patients.

Minimal change disease is the most common nephrotic glomerular disease in children.

Membranous nephropathy is the most common nephrotic glomerular disease in adults, especially those with lupus or Hepatitis B.

Membranoproliferative glomerulonephritis and IgA nephropathy both result in nephritic syndrome, characterized by microscopic hematuria.
A 25-year-old army recruit is brought into the emergency room after his first week of summer boot camp complaining of dizziness and decreased urination of 2 days duration. Physical exam shows a lethargic young male with sunken eyes and chapped lips. Vital signs are significant for a pulse of 120 bpm and a blood pressure of 100/60. Blood tests show a BUN of 45 mg/dL and a creatinine of 2 mg/dL. Urine analysis reveals a fractional excreted sodium of <1%. An abnormality in which of the following is the most likely cause for this patient’s condition?

a. Functional status of the urea cycle  
   b. Glomerular filtration rate (GFR)  
   c. Myocardial contractility  
   d. Protein content in the diet  
   e. Proximal tubule reabsorption

**Correct answer: B**

This patient’s clinical scenario is characteristic of acute renal failure secondary to hypovolemia. Acute renal failure (ARF) can manifest clinically as oliguria (100-400 ml in 24 hours) or anuria (<100 ml in 24 hours). ARF can be classified as pre-renal, intrinsic, or post-renal. Pre-renal ARF can be defined with labs showing BUN/Cr >20 and fractional excreted sodium <1%. Hypovolemia is a common cause of pre-renal ARF and can be identified clinically by signs of dehydration: dizziness, sunken eyes, dry mucous membranes, tachycardia, and hypotension. In this dehydrated patient, ARF is a result of decreased cardiac output from hypovolemia leading to a decrease in GFR (choice B).

Problems with the urea cycle (choice A) could explain the patient’s elevated BUN, but it would not explain his elevated creatinine and renal failure.

There is no evidence that this patient's myocardial contractility (choice C) is compromised. His tachycardia is likely due to decreased preload, secondary to hypovolemia. In fact, good myocardial contractility is allowing him to maintain a systolic blood pressure of 100 despite significant hypovolemia.

Dietary protein (choice D) can increase serum BUN, but it is not a potential etiology of ARF.

Dysfunction of proximal tubule reabsorption (choice E) is unlikely in this patient as demonstrated by his elevated BUN/Cr ratio and low fractional excreted sodium. Urea is reabsorbed by the proximal tubule while creatinine is not; thus, if the proximal tubule were damaged, the BUN/Cr ratio would be smaller. Additionally, the proximal tubule also reabsorbs sodium. Damaged proximal tubules would result in a larger fractional excreted sodium.
A 29-year-old G2 P1001 in her third trimester presents to the clinic for a routine check up. You take her vitals and note that her blood pressure is elevated to 150/95. On interview, she complains about some mild RUQ pain that she has had for the last few days. You confirm this on physical exam and also take note of facial puffiness. A urine dipstick in the office shows 3+ proteinuria. What are the laboratory features of the particular syndrome this patient has likely developed?

- Elevated BUN and elevated creatinine
- Elevated fasting glucose level and elevated liver enzymes
- Low platelet count and elevated liver enzymes
- Low platelet count and elevated magnesium level
- Low white blood cell count and elevated liver enzymes

**Correct answer:** C

The combination of hypertension, proteinuria and nondependent edema in a pregnant woman should immediately make you think about preeclampsia. The presence of right upper quadrant (RUQ) pain further suggests the development of HELLP syndrome, which occurs in about 10% of patients with preeclampsia. HELLP syndrome is Hemolysis, elevated LFTs, and low platelets. Thus, you definitely want to check the platelet count and the LFTs in this scenario.

WBC count would be helpful if you suspected an infection, but this is not the most likely diagnosis at this time.

Magnesium sulfate is used for seizure prophylaxis in women with preeclampsia (ie, to avoid eclampsia).

Renal failure can occur in preeclampsia, but checking renal function alone is not the best initial step.

Diabetic women are at an increased risk of developing preeclampsia if they become pregnant. However, measuring the fasting glucose level would not help diagnose preeclampsia or the HELLP syndrome.

Note: G2 P1001 tells us that the patient has been pregnant (G = gravida) twice. She has had 1 full term birth, 0 preterm births, 0 abortions (spontaneous or induced), and has 1 living child. Thus she is P, or para, 1-0-0-1. The mnemonic for this is F-P-A-L (Full term, Preterm, Abortions, Living children).
A 23-year-old woman has a progressive increase in her serum beta-human chorionic gonadotropin (beta-hCG) concentrations during an 8-week period. A hydatidiform mole is removed, but the beta-hCG concentration continues to increase. Which of the following is the most likely diagnosis?

a. Adrenal adenoma
b. Choriocarcinoma
c. Ectopic pregnancy
d. Pituitary insufficiency
e. A second noninvasive mole

**correct answer: B**

Steadily increasing beta-hCG concentration status post removal of hydatidiform mole is suggestive of malignant gestational trophoblastic disease (GTD). Choriocarcinoma (choice B) is a GTD that is commonly associated with increased beta-hCG concentrations and hydatidiform moles, making it the most likely diagnosis in this patient.

Adrenal adenoma (choice A) and pituitary insufficiency (choice D) are not commonly associated with either hydatidiform moles or increased concentrations of beta-hCG.

Ectopic pregnancy (choice C) or a second noninvasive mole (choice E) could be possible explanations for an increased beta-hCG concentration, but is much less likely than a malignant GTD in this clinical scenario.
A 24-year-old primigravid woman at 28 weeks' gestation has had nagging headaches, a puffy-looking face, and swollen legs for the past week. Her blood pressure is 180/95 mm Hg; it was within normal limits earlier in the pregnancy. Urinalysis shows a protein concentration of 0.6 g/dL. Which of the following is the most likely diagnosis?

a. Acute glomerulonephritis
b. Congestive heart failure
c. Eclampsia
d. Nephrotic syndrome
e. Preeclampsia

**Correct answer:** E

Proteinuria, edema, and hypertension after 20 weeks' gestation in a previously normotensive woman are pathognomonic for (E) preeclampsia. Preeclampsia occurs in approximately 5 to 8 percent in the United States, although it is mild in 75 percent of cases. This disease is associated with the HELLP syndrome (Hemolysis, Elevated LFTs, Low Platelets.) When the above presentation includes seizures, a diagnosis of (C) eclampsia is made. In this case, this woman's headaches are caused by hypertension, while her edema is secondary to the reduced plasma oncotic pressure following protein loss in the urine.

(A) Acute glomerulonephritis can cause hypertension, but is less likely to cause edema, while (D) Nephrotic syndrome causes edema without hypertension. (B) Congestive heart failure does not cause orbital edema.
A 55-year-old woman presents to the OB/Gyn clinic complaining of pain during sex, after which she experiences vaginal bleeding and foul discharge. She reports that she had multiple sexual partners in her youth, but has been monogamous for twenty years and has since had three children. She admits that has not seen a gynecologist in several years.

On physical exam, she is pale and her cervix is nodular, with what grossly appears to be some dysplasia. Which of the following organisms is the most likely cause of her condition?

a. Epstein Barr virus (EBV)
b. Human immunodeficiency virus (HIV)
c. Human papilloma virus (HPV)
d. Human T lymphotrophic virus, type 1 (HTLV-1)
e. Schistosoma hematobium

**Correct answer:** C

Human papilloma virus (HPV) is associated with cervical intraepithelial neoplasia (CIN) and cervical cancer. The risks for acquiring HPV and developing cervical cancer are:

1. Early age at first intercourse
2. Early age of first pregnancy
3. Multiple pregnancies
4. Multiple sexual partners
5. Cigarette smoking

Typically, CIN is discovered during routine Papaniculaou (Pap) smears, during which dysplastic cells are found. Any grossly suspicious lesion in the cervix should be biopsied.

Early-stage cervical cancer presents with irregular vaginal bleeding that occurs post-coital or at other intermenstrual times. More advanced cervical cancer may experience foul-smelling, watery vaginal discharge.
A 19-year-old sexually active female presents with a chief complaint of severe, stabbing lower abdominal pain. She also complains of foul-smelling vaginal discharge and pain with sex. On physical examination, she does not have a fever and complains of both abdominal and adnexal pain. Pelvic examination is difficult to perform secondary to pain, but her cervix appears erythematous with mucopurulent discharge coming from the os. When the bed upon which the patient is lying is jarred, she screams in pain. This patient is most likely suffering from an infection that originated in which of the following structures?

- a. Bladder
- b. Cervix
- c. Fallopian tubes
- d. Ovaries
- e. Rectum

**Correct answer:** B

This patient fits the clinical picture of acute salpingitis, or pelvic inflammatory disease (PID), the most common serious complication of sexually transmitted infections. PID generally begins as cervicitis (Choice B) caused by Neisseria gonorrhoeae and/or Chlamydia trachomatis. If untreated, the infection can ascend to the upper reproductive tract (Choice C and D). Although the definitive diagnosis is made by laparoscopy revealing pyosalpinx, the diagnosis of PID is made clinically with an elevated white blood cell count, fever, pelvic pain, cervical motion tenderness, and adnexal tenderness. The original infection most likely did not originate from either the bladder (Choice A) or rectum (Choice E).
A patient with a family history of gynecological cancer presents with concerns about her risk of developing one and various diagnostic strategies. Which of the following gynecological cancers is most likely to be staged clinically rather than surgically?

- a. Cervical
- b. Endometrial
- c. Ovarian
- d. Vaginal
- e. Vulvar

**Correct answer:** A

Cervical cancer is the only gynecological cancer that is clinically staged. Clinical staging involves predicting the amount of invasion into adjacent structures and metastatic involvement. Diagnostic tools include exam under anesthesia, chest X-ray, cystoscopy, proctoscopy, intravenous pyelogram, and barium enema. MRI and CT can be used to define the extent of the disease but not to determine the stage. Intraoperative findings or disease progression does not alter stage assignment.

Endometrial cancer (Choice B) is surgically staged with total abdominal hysterectomy and bilateral salpingo-oophorectomy (TAHBSO) as it relies on pathologic confirmation. During surgery, the uterus should be opened to determine the depth of myometrial invasion. Most endometrial cancers are diagnosed at stage I.

Ovarian cancer (Choice C) is surgically staged using TAHBSO, omentectomy, peritoneal washing and cytology, Pap smear of the diaphragm, and sampling of pelvic and para-aortic lymph nodes. Nearly 75% of patients present with stage III or IV cancers.

Vaginal cancer (Choice D) is surgically staged based on tumor size and invasiveness, nodal involvement, and distant metastases. The most important prognostic factor is the number of positive inguinal lymph nodes.

Vulvar cancer (Choice E) is surgically staged similarly to vaginal cancer.
Neonatal respiratory distress syndrome (also known as hyaline membrane disease) is a major cause of death in premature infants and is the result of inadequate surfactant production in the lungs. Which of the following can be used to determine the maturity of fetal lungs in utero?

a. Amniotic fluid analysis  
b. Chorionic villus sampling  
c. Fetal heart rate monitor  
d. Karyotype analysis  
e. Ultrasound

**Correct answer:** A

Amniotic fluid can be analyzed to determine the lecithin to sphingomyelin ratio. A ratio > 2 is rarely associated with Neonatal respiratory distress syndrome (NRDS). In NRDS the ratio is usually < 1.5.

The other tests are not the test of choice for determining fetal lung maturity.
A 60-year-old man has a 5-day history of productive cough and shortness of breath with exertion. In addition to a normal left lung base, examination of the chest in the area of the right lung base shows: Breath sounds bronchial, Percussion note dull, Tactile fremitus increased, Adventitious sounds, and crackles. Which of the following is the most likely diagnosis?

- a. Asthmatic bronchitis
- b. Bullous emphysema
- c. Chronic bronchitis
- d. Congestive heart failure
- e. Lobar pneumonia

**Correct answer:** E

This patient has a productive cough with unilateral signs of alveolar consolidation as demonstrated by right-sided bronchial breath sounds and increased tactile fremitus. In this clinical setting, lobar pneumonia (choice E) is the most likely diagnosis.

Asthmatic bronchitis (choice A) is characterized by a nocturnal cough, episodic expiratory wheezing (inspiratory as well when severe), and decreased breath sounds due to air-trapping.

Bullous emphysema (choice B) is characterized by progressive dyspnea, diminished breath sounds, and decreased tactile fremitus due to air trapping. A productive cough is not commonly associated with this condition.

Chronic bronchitis (choice C) is defined as a productive cough for at least 3 months for 2 consecutive years.

Congestive heart failure (choice D) can lead to bilateral pleural effusions as the pulmonary capillary hydrostatic pressure exceeds the capillary oncotic pressure and fluid moves into the pleural space. Clinically, CHF can be characterized by bilateral decreased breath sounds, crackles at both lung bases, decreased tactile fremitus bilaterally. Fremitus is decreased or absent when the transmission of vibrations from the larynx to the surface of the chest is impaired. Etiologies include obstructed bronchus, COPD, separation of the pleural surfaces by fluid, fibrosis, infiltrating tumor, or a very thick chest wall.
A previously healthy, tall, and slender 19-year-old woman has the sudden onset of right-sided chest pain followed by progressive dyspnea. Ten hours later, an x-ray of the chest shows a collapsed right lung and air in the right pleural space. Which of the following is the most likely underlying condition?

a. Bronchiectasis  
b. Lung abscess  
c. Panacinar emphysema  
d. Pulmonary sequestration  
e. Subpleural blebs

**Correct answer:** E

Primary spontaneous pneumothorax presents with acute chest pain and shortness of breath. It is believed to result from rupture of a subpleural bleb (choice E), which is usually located in the apex of the lung. Blebs can be found in more than 75% of patients undergoing thoracoscopy for treatment of primary spontaneous pneumothorax. Also, patients with a spontaneous pneumothorax tend to be thinner and taller on average as is the case with this patient. An important distinction of the correct answer from the other answer choices is that it is the only choice that most likely will have an acute presentation in adulthood.

Bronchiectasis (choice A) and lung abscess (choice B) are both caused by infectious processes that would be unlikely in a previously healthy patient.

Panacinar emphysema (choice C) and pulmonary sequestration (choice D) would both most likely present much earlier in life. Panacinar emphysema is most commonly a result of alpha-1-protease inhibitor deficiency and pulmonary sequestration is a congenital malformation of the lower respiratory tract which results in a nonfunctioning mass of lung tissue.
A homeless man presents to the ER with multiple bruises, swollen gums, and petechiae. His diet for the past two years has consisted solely of red meat and diet coke. What is the likely diagnosis?

a. Beriberi  
b. Pellagra  
c. Rickets  
d. Scurvy  
e. Wernicke-Korsakoff Syndrome

**Correct answer:** D  
The patient's presentation is typical of Vitamin C deficiency and scurvy. Beriberi (thiamine deficiency) has neuropathic consequences. Pellagra produces the three Ds (dementia, diarrhea, and dermatitis). Rickets is caused by Vitamin D deficiency and leads to weak bones/easy fracturing. Wernicke-Korsakoff syndrome is also caused by severe thiamine deficiency and has numerous neurological & gait abnormalities.
A 72-year-old man who is a retired construction worker comes to the physician because he has had a lesion on his face for 3 months. Physical examination shows a 6-mm, red, ulcerated lesion with heaped borders. A biopsy specimen of the lesion shows atypical, dysplastic keratinocytes within the epidermis and dermis. Which of the following is the most likely diagnosis?

a. Actinic keratoses
b. Discoid lupus erythematosus
c. Melanoma
d. Mycosis fungoides
e. Squamous cell carcinoma

**Correct answer:** E

Skin cancer comes in three major forms - squamous cell carcinoma (SCC), basal cell carcinoma (BCC), and melanoma. This patient's presentation and histology suggest squamous cell carcinoma. In general, a lesion described as ulcerated or with heaped borders is cancerous until proven otherwise. The atypical and dysplastic keratinocytes indicate a squamous cell origin.

Actinic keratosis is a thick, scaly lesion that a predecessor lesion to SCC. It would be fair to consider in this patient except that the clinical presentation describes a lesion that has progressed much further.

Melanoma is a neoplasm of melanocytes. Melanoma should be associated with ABCDE - Asymmetry, irregular Borders, multiple Colors, Diameter greater than 5mm, and Evolution of a pre-existing nevus. Melanoma is more common in younger people.

Mycosis fungoides is a presentation of cutaneous T-cell lymphoma; this patient's histology is not suggestive of such a rare diagnosis.
A 78-year-old male experiences a hypertensive emergency and is given sodium nitroprusside intravenously. This patient's nitric oxide levels can be monitored by which of the following laboratory tests?

- a. Methemoglobin levels
- b. Plasma nitroprusside concentration
- c. Serum thiocyanate levels
- d. Urinary cyanide concentration
- e. Urinary nitroprusside concentration

**Correct answer:** C

Nitroprusside is non-enzymatically transformed to cyanide to form cyanomethemoglobin. The cyanide is then metabolized to thiocyanate in the plasma. This assay can assess the effectiveness of therapy. Plasma nitroprusside concentration is incorrect because the half life of the drug is usually too short to make this a meaningful test, and the other choices are biochemically and physiologically not feasible.
A 30-year-old woman with a 1-week history of severe diarrhea feels dizzy when she stands up. Blood pressure (while supine) is 112/76 mm Hg with a pulse of 88/min; blood pressure (while standing) is 80/60 mm Hg with a pulse of 120/min. In addition to controlling her diarrhea, the most appropriate initial therapy is intravenous administration of which of the following?

a. Desmopressin  
b. 5% Dextrose in water  
c. Fresh frozen plasma  
d. 0.9% Saline  
e. Methoxamine

**correct answer:** D

This patient is exhibiting signs and symptoms of orthostatic (postural) hypotension in the setting of decreased intravascular volume secondary to prolonged diarrhea. Orthostatic hypotension is diagnosed when, within 2-5 minutes of standing, one or more of the following are present: (1) 20mmHg or more fall in systolic pressure; (2) 10mmHg or more fall in diastolic pressure; (3) symptoms of cerebral hypoperfusion. There are many etiologies of orthostatic hypotension which include decreased intravascular volume, autonomic insufficiency, alcohol, medications, and aging. The patient’s prolonged diarrhea has left her in a volume depleted state, causing the apparent signs and symptoms. The best initial therapy for this patient is to replete her intravascular volume with intravenous fluids that are as similar to her plasma as possible. The patient has lost both water and electrolytes in her diarrhea, both of which need to be replaced. Of the answer choices 0.9% Saline (choice D) both resembles plasma the most and is cost-effective. 0.9% Saline is also known as normal saline and is comprised of 154mEq of sodium and 154mEq of chloride.

Desmopressin (choice A) is a synthetic arginine vasopressin analogue that has antidiuretic effects. It can be used to treat patients with central diabetes insipidus. This patient most likely has high levels of ADH in her volume deplete state and would benefit more with IV hydration.

5% Dextrose in water (choice B) is a type of IV fluid; however, it is not as physiologically similar to plasma as 0.9% Saline. 5% dextrose in water is essentially water with dextrose without any electrolytes. Furthermore, as the dextrose will be rapidly metabolized, the water supplied distributes across both the extracellular and intracellular fluid compartments. This leads to a reduced amount of intravascular volume repletion relative to supplying saline.

Fresh frozen plasma (choice C) is a blood product with strict indications, only given to patients to replace clotting factors, not to correct volume status.

Methoxamine (choice E) is an alpha-1-adrenergic receptor agonist that induces vasoconstriction. This patient would benefit more from correcting volume status with IV fluids than with a vasoconstrictor.
A 65 year old man with a history of essential hypertension presents with bilateral, non-tender leg swelling for 2 weeks. His hypertension was diagnosed three weeks ago and he is currently taking only one anti-hypertensive. He has no signs or symptoms of congestive heart failure and no edema in other body parts. Which of the following medications is most likely to be responsible for his edema?

a. Hydrochlorothiazide  
b. Furosemide  
c. Enalapril  
d. Metoprolol  
e. Amlodipine

**Correct answer: E**

Amlodipine is a calcium channel blocker. It works by inhibiting transmembrane influx of calcium in peripheral and cardiac vascular smooth muscle. The result is reduced vascular tone, which can relieve coronary vasospasm (angina) and decrease blood pressure. Vasodilation in peripheral tissues may give rise to vascular leakage. The side effect of lower extremity edema is a common indication for discontinuation of CCB.

HCTZ is a thiazide diuretic that is a first line treatment for essential hypertension. Its main effect is to reduce effective arterial volume via diuresis. Common side effects are hypercalcemia and electrolyte imbalances.

Furosemide is used to relieve volume overload, usually pulmonary edema, in congestive heart failure. It is not a common treatment for isolated essential hypertension. Common side effects are hyponatremia and hypocalcemia, as well as frequent need to urinate.

Enalapril is an angiotensin converting enzyme inhibitor. ACE inhibitors reduce hypertension by blunting the renin-angiotensin system, thus reducing the kidney's holding on to sodium. The most common side effect of ACE inhibitors is coughing, followed by angioedema (e.g. swollen lips).

Metoprolol is selective beta-1-adrenergic blocker, hence the target tissue is cardiac muscle. Common side effects include bradycardia, exercise-induced syncope, and worsening of obstructive lung disease (though less so than a nonselective beta blocker like propranolol).
A 25-year-old woman presents with 6 months of lethargy, sensitivity to cold temperatures, and an elevated serum TSH. What is the most appropriate management for this patient?

a. Fluoxetine  
b. Hydrocortisone  
c. Levothyroxine  
d. Propylthiouracil  
e. Radioactive iodine treatment

**Correct answer:** C

This woman complains of symptoms of hypothyroidism with an elevated TSH. Levothyroxine, a synthetic form of thyroid hormone, is the most appropriate choice.

(A) Fluoxetine is an SSRI could be used for depression. Serum TSH would be normal in depression.

(B) Hydrocortisone is a steroid that could be used for Addison’s Disease (chronic adrenal insufficiency). Serum TSH should be normal in Addison’s.

(D) Propylthiouracil and (E) radioactive iodine treatment are used in hyperthyroidism. Serum TSH would be low in hyperthyroidism. Propylthiouracil inhibits the enzyme thyroperoxidase, thus resulting in decreased thyroxine production. It also inhibits the peripheral deiodination of T4 to T3 (T3 is more active). In radioactive iodine treatment, there is thyroid uptake of the radioactive isotope iodine-131, which results in cytotoxic radiation delivered to some thyroid cells.
A 30-year-old woman known to have type 1 diabetes presents to the emergency room with a productive cough, fever, and polyuria for 4 days and progressive lethargy and confusion for the last day. She complains of abdominal pain and nausea with vomiting. Her breath smells fruity, her serum glucose is 600 mg/dL, and her serum potassium is 5.5. Initial management of this patient will likely include IV fluids, insulin, and which of the following?

- a. Calcium
- b. Cortisone
- c. Glucagon
- d. Kayexalate
- e. Potassium

**Correct answer:** E

This woman is likely in diabetic ketoacidosis. She needs urgent IV fluids, insulin, and potassium. Despite a mildly elevated serum potassium her total body potassium stores are low. Administration of insulin will drive potassium into cells and decrease her serum potassium.
A 50-year-old man being treated for increased serum LDL concentration has muscle pain that has recently become more severe, especially with exercise. He is most likely to be taking a drug with which of the following mechanisms of action?

- a. Binding bile acids
- b. Decreasing binding of bile acids
- c. Decreasing production of LDL
- d. Decreasing the activity of lipoprotein lipase
- e. Inhibiting the activity of 3-hydroxy-3-methylglutaryl (HMG) CoA reductase

**Correct answer:** E

This patient is most likely taking a statin. Statins inhibit the activity of HMG CoA reductase (choice E). One of the major side effects reported from statin use is myalgia. This is an important presenting symptom because continued use can result in rhabdomyolysis and acute renal failure. Decreasing the production of LDL (choice C) is an effect of statins, but not the direct action.

Bile acid binding resins (choice A) such as cholestyramine do not cause myalgias.

Decreasing the binding of bile acids (choice B) and decreasing the activity of lipoprotein lipase (choice D) are not current methods for lowering serum cholesterol.
Warfarin is administered to a 56-year-old man following placement of a prosthetic cardiac valve. The warfarin dosage is adjusted to maintain an INR of 2.5. Subsequently, trimethoprim-sulfamethoxazole therapy is begun for a recurring urinary tract infection. In addition to monitoring prothrombin time, which of the following actions should the physician take to maintain adequate anticoagulation?

a. Begin therapy with vitamin K
b. Increase the dosage of warfarin
c. Make no alterations in the dosage of warfarin
d. Decrease the dosage of warfarin
e. Stop the warfarin and change to low-dose aspirin

**Correct answer:** D

Trimethoprim-sulfamethoxazole is a sulfa-based drug. Sulfonamides are known inhibitors of the P-450 enzymatic system, and warfarin is primarily metabolized by CYP450:2C9. Trimethoprim-sulfamethoxazole would be expected to increase the serum levels of warfarin and push the INR to super-therapeutic levels with the increased risk of bleeding. Therefore, it would be recommended to decrease the dosage of warfarin (choice D).

Beginning therapy with vitamin K (choice A) is the treatment for warfarin overdose, which this patient does not have. Additionally, giving vitamin K will drop the INR to sub-therapeutic levels which will place this patient with a newly placed prosthetic valve at risk for thromboembolisms.

Increasing the dosage of warfarin (choice B) would not be recommended in this patient because trimethoprim-sulfamethoxazole will already increase the serum levels of warfarin. Further increasing the dosage of warfarin will definitely place this patient at risk for bleeding.

Making no alterations in the dosage of warfarin (choice C) is not recommended because of the risk of bleeding with a supra-therapeutic INR as a result from inhibited warfarin metabolism.

Stopping the warfarin and changing to low-dose aspirin (choice E) is not recommended because this patient with a newly placed prosthetic valve still requires anti-coagulation to prevent thromboembolisms. Additionally, aspirin has not been shown to be adequately effective in anti-coagulating patients with prosthetic valves.
A 26-year-old man who is HIV positive has a CD4+ T-lymphocyte count of 250/mm3 (N≥500). After 5 weeks of therapy with two nucleoside reverse transcriptase inhibitors and a protease inhibitor, he feels weak and is easily fatigued. His hemoglobin concentration has decreased from 12.8 g/dL to 8.2 g/dL. Which of the following is the most likely cause of the anemia in this patient?

a. Decreased formation of erythrocytes
b. Folic acid deficiency
c. Increased formation of erythrocyte antibodies
d. Increased fragility of erythrocytes
e. Iron deficiency

**Correct answer:** A

The commonly-used HIV medications have well-known side effects. In particular, zidovudine (AZT), the commonly-used nucleoside reverse transcriptase inhibitor (NRTI), may cause anemia through (A) decreased formation of erythrocytes. Approximately 1% of patients experience this adverse effect after 2-4 weeks of taking the medication. Other significant side effects of AZT are headache (63%), malaise (53%), nausea (51%), anorexia (20%), vomiting (17%), constipation (6%), and granulocytopenia (2%), anemia (1%).

(B) Folic acid deficiency and (E) iron deficiency may, respectively, cause macrocytic and microcytic anemia, but they are not in and of themselves related to HIV medications. Similarly, these medications do not affect (C) the formation of erythrocyte antibodies or (D) the fragility of erythrocytes.
The thrombolytic effect of tissue plasminogen activator depends on which of the following?

a. Circulating heparin
b. Concomitant therapy with high doses of aspirin
c. Factor II (prothrombin) concentration
d. Fibrinogen concentration
e. Presence of fibrin at the site of the thrombus

**Correct answer:** E

Activated plasminogen (plasmin) degrades fibrin to fibrin split products.

(A) Heparin activates antithrombin III, which decreases the activity of IIa and Xa.
(B) Aspirin inhibits cyclooxygenase and thereby prostaglandin production by platelets.
(C) Factor II (prothrombin) is the inactive form of IIa (thrombin).
(D) TPA action is not dependent on fibrinogen concentration.
An 8-year-old boy has had a temperature of 38.8°C along with an upper respiratory tract infection over the past two days. His mother gives him medication to bring down his fever. Two days later, the boy becomes increasingly confused and lethargic, and starts vomiting heavily. Physical exam reveals a slightly enlarged and firm liver. His eyes are anicteric. Which of the following medications is likely responsible for this boy’s condition?

a. Acetaminophen  
b. Acetylsalicylic acid  
c. Ibuprofen  
d. Indomethacin  
e. Naproxen

**correct answer: B**

Acetylsalicylic acid (choice B) is a non-selective cyclooxygenase (COX) inhibitor, which has anti-inflammatory, anti-pyretic, anti-platelet, and analgesic effects. One notable adverse effect is Reye’s syndrome, which is a potentially fatal condition seen in children under 15 years who take acetylsalicylic acid after an upper respiratory infection. The condition manifests as rapidly progressive liver failure and encephalopathy. Acetaminophen (choice A) also inhibits the COX enzyme, but has no anti-inflammatory or anti-platelet effects. Ibuprofen (choice C), indomethacin (choice D), and naproxen (choice E) are other non-selective COX inhibitors.
Which of the following is a contraindication for usage of the drug cyclophosphamide?

a. Liver disease  
b. Lung disease  
c. Heart disease  
d. Hypertension  
e. Diabetes

**Correct answer:** A

Cyclophosphamide is a nitrogen mustard alkylating agent that crosslinks DNA, making it useful in cancer treatment (such as leukemias). It is a pro-drug that undergoes bioactivation in the liver, and therefore advance hepatic disease will decrease its therapeutic activity.
A lacrosse player develops athlete's foot (tinea pedis). A drug with which the following mechanisms would be most effective for this patient?

- a. Binding to hemaglutinin
- b. Blockade of mycolic acid incorporation into cell wall
- c. Inhibition of ergosterol synthesis
- d. Inhibition of reverse transcriptase
- e. Inhibition of transcription

**Correct answer:** C

Tinea pedis is a fungus and therefore does not have mycolic acids (B), reverse transcriptase (D), or hemaglutinin (A). A transcription inhibitor (E) would also affect the patient's normal cells, resulting in terrible side effects. Therefore, inhibition of ergosterol synthesis (C) is the most effective therapy.
A 22-year-old college student is brought to the emergency room by his parents because he's been behaving unusually for the last 6 months. His parents report he's been having frequent angry outbursts and tends to spend much more time alone in his room while at home. Upon questioning the patient reports that he has spent more time alone recently because the FBI is spying on him and he's not sure who he can trust. This patient could likely benefit from a medication that primarily acts on receptors for which neurotransmitter?

a. Acetylcholine
b. Dopamine
c. GABA
d. NMDA
e. Norepinephrine

**Correct answer: B**

This patient is likely to be diagnosed with schizophrenia. Classic antipsychotics (neuroleptics) act mainly on dopamine receptors. Atypical antipsychotics act on both 5-HT and dopamine receptors.
Oral administration of grapefruit juice increases the bioavailability of a variety of drugs, including felodipine, midazolam, saquinavir, and verapamil. After oral administration of any one of these drugs, which of the following is most likely to be increased in the presence of grapefruit juice?

- a. Hepatic CYP3A4 activity
- b. Intestinal drug metabolism
- c. Peak serum concentration
- d. Total body clearance
- e. Volume of distribution

**Correct answer:** C

Recent studies have demonstrated that grapefruit juice contains chemicals called furanocoumarins which have been shown to inhibit drug metabolism by interfering with the hepatic cytochrome P450 system, especially CYP3A4. Inhibiting a drug's hepatic metabolism can lead to higher peak serum concentrations (choice C) of that drug.

Hepatic CYP3A4 activity (choice A) would be decreased by the furanocoumarins in grapefruit juice.

Intestinal drug metabolism (choice B) should not be affected because grapefruit juice inhibits hepatic drug metabolism.

Total body clearance (choice D) and volume of distribution (choice E) of the aforementioned drugs should both decrease. Volume of distribution is the ratio of the amount of drug in the body versus the plasma drug concentration. If the plasma drug concentration increases because drug metabolism is decreased, then volume of distribution will decrease. Clearance is the ratio of the rate of elimination of a drug to the plasma drug concentration. If the plasma drug concentration increases, then clearance will decrease.
Four drugs produce the same therapeutic effect by the same mechanism, but each has a unique toxicity. When used at a maximally effective therapeutic dose, the drug with which of the following sets of characteristics is most likely to have the highest incidence of toxicity?

a. Median Effective Dose 0.5x, Toxic Dose/Effective Dose 2.8
b. Median Effective Dose 1x, Toxic Dose/Effective Dose 2.1
c. Median Effective Dose 5x, Toxic Dose/Effective Dose 4.0
d. Median Effective Dose 33x, Toxic Dose/Effective Dose 3.0
e. N/A

**Correct Answer: B**

The closer the toxic dose is to the effective dose, the narrower the therapeutic window. Therefore, the ratio of toxic/effective dose is what matters. If it is closer to 1, it has the highest risk of toxicity at effective doses, regardless of what the median effective dose actually is. Examples of drugs that have narrow therapeutic windows include lithium and digoxin.
A 45-year-old woman who is being treated for hypertension and hypercholesterolemia develops diffuse muscle pain and weakness. Her serum creatine kinase activity is increased. Which of the following drugs is most likely to have caused this clinical picture?

a. Captopril
b. Hydrochlorothiazide
c. Lovastatin
d. Nicotinic acid
e. Propranolol

**Correct answer:** C

This patient may be developing rhabdomyolysis - a very serious complication involving muscle necrosis and release of cellular contents into the bloodstream. Clinical signs include diffuse muscle pain, increased serum creatine kinase, elevated serum potassium, and acute renal failure - indicated by a rapidly rising creatinine level. Although rhabdomyolysis is a rare side effect, it has been associated with the administration of statins, such as lovastatin, especially when taken in combination with cyclosporine or fibrates. None of the other drugs listed above has been associated with rhabdomyolysis.
A 32-year-old man is brought to the emergency department because of confusion, wheezing, vomiting, and diarrhea for the past 6 hours. He is sweating and salivating profusely. There is generalized muscle weakness. Which of the following substances is the most likely cause of these findings?

a. Glutethimide  
b. Heroin  
c. Jimson weed (belladonna alkaloids)  
d. Parathion  
e. Phencyclidine (PCP)

**correct answer:** D

This patient has been poisoned by the organophosphate parathion. Organophosphates inhibit acetylcholinesterase (AChE), leading to toxic build-up of acetylcholine (ACh) and resulting in the symptoms listed above. Remember the mnemonic DUMBELS associated with cholinergic toxicity: Defecatioin, Urination, Miosis, Bronchorrhea/Bronchospasm/Bradycardia, Emesis, Lacrimation, Salivation.
A 55-year-old woman with breast cancer develops shortness of breath and poor exercise tolerance while being treated with doxorubicin. Which of the following is the most likely cause of these symptoms?

a. Dilated cardiomyopathy  
b. Obstructive pulmonary disease  
c. Pulmonary hypertension  
d. Restrictive cardiomyopathy  
e. Restrictive pulmonary disease

**Correct answer:** A

Doxorubicin has a well known side-effect profile of (A) dilated cardiomyopathy, with dose-related CHF and transient ECG abnormalities occurring in over 10% of patients. Other side effects include leukopenia (75%) nausea and vomiting (21-55%). It is not as closely associated with restrictive cardiomyopathy or pulmonary disease. The chemotherapeutic agent bleomycin is better known for causing pulmonary toxicity, particularly pulmonary fibrosis.
A 12-year-old boy has a pruritic, coin-shaped, scaly erythematous lesion with a raised border and central clearing on his right arm. Potassium hydroxide preparation results are consistent with tinea corporis. A drug with which of the following mechanisms of action is most likely to be effective?

- a. Blockade of ergosterol synthesis
- b. Inhibition of DNA synthesis
- c. Inhibition of ribosomal protein synthesis
- d. Interference with mycolic acid synthesis
- e. Irreversible binding to DNA-dependent RNA polymerase

**Correct answer:** A

Tinea corporis or ringworm is a fungal infection. An effective treatment for this infection is a topical azole that acts by blocking ergosterol synthesis and thereby increasing fungal membrane permeability. Miconazole and clotimazole are two azoles available in topical solutions that can be used to treat this infection.

Inhibition of DNA synthesis is the mechanism of action of flucytosine, which converts to fluorouracil and competes with uracil. This drug can be used to treat systemic fungal infections, but the side effects of nausea, vomiting, diarrhea, and bone marrow suppression limit its use.

Inhibition of ribosomal protein synthesis is the mechanism of action of various antibacterial agents including aminoglycosides, tetracyclines, chloramphenicol, erythromycin, Lincomycin, and Clindamycin. These drugs are used to treat various bacterial infections, but they are not used to treat fungal infections.

Interference with mycolic acid synthesis is how Isoniazid works. Isoniazid is a drug used for tuberculosis prophylaxis and in combination with other drugs for therapy for active tuberculosis. Isoniazid is not used to treat fungal infections.

Rifampin is another drug used in combination with Isoniazid for active tuberculosis therapy. It works by irreversibly binding to and inhibition of DNA-dependent RNA polymerase. Rifampin also is used for meningococcal prophylaxis, but it is not used to treat fungal infections.
An otherwise healthy 55-year-old Asian American man is given isoniazid and vitamin B6 (pyridoxine) after conversion of his PPD skin test. An x-ray of the chest shows no abnormalities. Four weeks later, he develops abdominal pain and jaundice. Which of the following is the most likely explanation?

a. Hepatic tuberculosis
b. Hepatitis B
c. Isoniazid-induced hepatitis
d. Pyridoxine-induced cholecystitis
e. Tuberculous pancreatitis

correct answer: C

Hepatotoxicity is a well known side effect of isoniazid. Pyridoxine is given with isoniazid to help prevent B6 deficiency and neurotoxicity. The proximity of this man's symptoms to starting isoniazid, normal chest x-ray, and lack of exposure make tuberculosis and hepatitis B unlikely. Cholecystitis is not a well-known toxicity of pyridoxine and jaundice is not a typical feature of cholecystitis.
A 25-year-old professional cyclist is diagnosed with testicular cancer and undergoes chemotherapy. One month after beginning treatment, the patient complains of difficulty hearing. Which of the following chemotherapeutic agents is most likely responsible for this side effect?

a. Bleomycin  
b. Busulfan  
c. Cisplatin  
d. Cytarabine  
e. Doxorubicin

**correct answer:** C

This question is testing your knowledge of the major toxicities of chemotherapeutic agents.

Cisplatin (choice C), an alkylating-like agent, crosslinks DNA thereby interfering with mitosis. It is known to cause nephrotoxicity and ototoxicity; both side effects seem to be related to the production of reactive oxygen species in specific tissues, leading to cell death.

Bleomycin (choice A) induces the formation of free radicals, which causes breaks in DNA strands. It is known to cause pulmonary fibrosis, skin changes and occasionally bone marrow suppression.

Busulfan (choice B) is a DNA alkylator and is known to cause pulmonary fibrosis and skin changes, including hyperpigmentation.

Cytarabine (choice D) inhibits DNA polymerase and causes megaloblastic anemia and pancytopenia.

Doxorubicin (choice E) generates free radicals and intercalates in DNA, creating breaks in the DNA. It causes cardiotoxicity, hair loss and bone marrow suppression.
A 22-year-old college student presents to the ER complaining of difficulty speaking and the inability to swallow. He says he recently returned from spending Thanksgiving at his grandparents’ house, and yesterday had eaten a family favorite, home-canned asparagus prepared by his grandmother. He denies other complaints and was in his usual state of excellent health before the onset of symptoms. Which of the following is the most appropriate treatment?

- Administration of trivalent botulinum antitoxin
- Administration of staphyloccocal enterotoxin antiserum
- IV Amphotericin B
- Immunization with s. aureus enterotoxin toxoid
- PO ampicillin

**Correct answer: A**

The patient has symptoms of botulism, caused by exposure to Clostridium botulinum toxin, which may occur following ingestion of improperly canned low-acid content vegetables such as asparagus, corn, beets, and green beans. The best treatment is early administration of botulinum antitoxin containing antibodies to the three common types of C. botulinum toxin known to cause disease in humans (A,B,E). Staphyloccocal enterotoxin antiserum (B) may be appropriate in patients suffering from Staphylococcus aureus induced food poisoning, but this patient is not. The other choices would provide no benefit to this patient.
A 30-year-old man with peptic ulcer disease suddenly develops pain, redness, and swelling of his right first metatarsophalangeal joint. There is no history of injury. Serum uric acid concentration is 8 mg/dL. Examination of joint aspirate shows birefringent crystals. Which of the following drugs is most appropriate to treat the acute symptoms in this patient?

- a. Allopurinol
- b. Colchicine
- c. Morphine
- d. Probenecid
- e. Sulfinpyrazone

**Correct answer:** B
Colchicine is effective in acute gout. It inhibits polymerization of microtubules and leukocyte chemotaxis.

(A) Allopurinol, (D) probenecid, and (E) sulfinpyrazone should be avoided during an acute flair because rapid changes in serum uric acid concentrations can precipitate crystallization of uric acid. Allopurinol inhibits uric acid production by inhibiting xanthene oxidase and probenecid inhibits uric acid reabsorption in the kidney. Sulfinpyrazone's mechanism of action is similar to probenecid.

(C) Morphine is a nonspecific treatment choice for gout. Colchicine is a better first choice.
A 25-year-old man is started on clozapine for schizophrenia, paranoid type. He had been unsuccessfully treated for the past 2 months with haloperidol. The patient should be monitored for which of the following adverse effects?

a. Decreased erythrocyte count
b. Decreased leukocyte count
c. Decreased platelet count
d. Increased eosinophil count
e. Increased hemolysis

_correct answer: B_

Agranulocytosis (Answer B) is a rare adverse effect of all antipsychotics. The RBC (Answer A and E), platelet (Answer C) and eosinophil (Answer D) lines are not known to be affected by clozapine.

Clozapine is a (new-generation) atypical anti-psychotic that blocks 5-HT2 and dopamine receptors. Unlike the traditional antipsychotics, it is useful for treatment of the negative symptoms of schizophrenia. Its major side effect is agranulocytosis, which requires WBC monitoring.

Haloperidol is a high-potency dopamine blocker used to treat psychosis. Drugs in this class are more selective for dopamine receptors compared to low-potency antipsychotics such as chlorpromazine or thioridazine, hence haloperidol is the drug of choice to avoid cross-reactivity at muscarinic, adrenergic, and histaminic receptors. Long-term adverse effects of haloperidol include parkinsonian features and tardive dyskinesia secondary to blockade of the nigrostriatal pathway.
A 17 year old male has had numerous episodes of alternating stiffening and relaxing movements involving the entire body. On separate occasions, he was also noted to have episodes of staring blankly that last for about a minute. After prescribing the appropriate pharmacologic treatment, which of the following is most important to monitor as follow-up in this patient?

a. BUN and creatinine
b. Complete Blood Count (CBC)
c. Liver Function Tests (LFTs)
d. Pulmonary Function Tests (PFTs)
e. Serum electrolytes

**Correct answer: C**

This patient presented with tonic-clonic and absence seizures. Valproic acid is the only commonly used anti-epileptic drug that treats both types of seizures. The major toxicity of valproic acid is hepatotoxicity, for which LFTs must be monitored regularly (choice C).
A patient presents to your office complaining of a recent inability to have an orgasm when engaging in sexual activities with his wife. He recently started taking fluoxetine to control longstanding depression. Which of the following medications might you consider switching him to in order to best decrease his risk for sexual side effects?

- a. mirtazapine
- b. imipramine
- c. bupropion
- d. trazodone
- e. citalopram

**Correct answer:** C

Bupropion is an antidepressant medication that is known to be least likely to cause sexual side effects. It works by blocking the reuptake of dopamine and norepinephrine. Interestingly, it is also used to aid in smoking cessation, as it acts as a nicotinic receptor antagonist.
A 5-year-old boy with leukemia is treated with chemotherapy that includes ifosfamide. 48 hours after treatment, he presents with painless gross hematuria. What agent could have been given to prevent this side effect?

a. Allopurinol  
b. Mesna  
c. Misoprostol  
d. Pralidoxime (2-PAM)  
e. Thiosulfate

**Correct answer:** B

Alkylating chemotherapy agents such as ifosfamide and cyclophosphamide may degrade to the metabolite acrolein, which can damage the bladder, producing the symptoms described.

Mesna binds to acrolein and prevents it from causing this damage.

Allopurinol inhibits xanthine oxidase and may reduce the probability of developing urate kidney stones. Kidney stones would present with pain.

Misoprostol (PGE1) is used to prevent the development of ulcers in patients taking NSAIDs long term.

Pralidoxime is used to reverse the phosphorylation of acetylcholinesterase in patients that have been exposed to pesticides or nerve gases.

Thiosulfate scavenges cyanide ions in patients with cyanide poisoning.
A 31-year-old man presents to the clinic for his yearly check-up and is found to have a blood pressure of 158/94. At a re-check two weeks later, his blood pressure is still elevated even though he has no risk factors for the development of hypertension. Further evaluation reveals a potassium level of 3.3 meq/L. Proper medical therapy for this man’s most likely condition would involve which of the following mechanisms?

- a. Agonism of the aldosterone receptor in the distal renal tubules
- b. Antagonism of the aldosterone receptor in the distal renal tubules
- c. Inhibition of sodium and chloride reabsorption in the ascending loop of Henle and distal renal tubule
- d. Inhibition of sodium and water reabsorption in the proximal tubule and the loop of Henle
- e. Inhibition of carbonic anhydrase activity

**Correct answer:** B

The correct answer is B, antagonism of the aldosterone receptor in the distal renal tubules. This man most likely has primary hyperaldosteronism given the hypertension accompanied by hypokalemia. The goal of medical therapy is to normalize the serum potassium level and the blood pressure, and reverse the effects of hyperaldosteronism on the heart since hyperaldosteronism is associated with increased risk of cardiovascular disease and morbidity. Thus, therapy should be aimed at antagonism of mineralocorticoid receptors.

The other choices include the mechanisms of action for other diuretics. These drugs are often used for hypertensive patients, but they are not the best medications to use for patients with primary aldosteronism. Inhibition of sodium and chloride reabsorption in the loop of Henle and distal renal tubule (Answer C) is achieved with loop diuretics including furosemide. Inhibition of sodium and chloride reabsorption in the proximal tubule and loop of Henle is the mechanism of action of mannitol, an osmotic diuretic (Answer D). Inhibition of carbonic anhydrase activity is how acetazolamide works (Answer E).

Agonism of the aldosterone receptor in the distal renal tubules is merely how aldosterone works and is a likely mechanism behind this patient's hypertension, but it result in worsening of his hypertension and hypokalemia instead of improvement.
Looking in his medicine cabinet, a 45-year-old man with fever, nasal congestion, fatigue, and arthralgias finds a drug that was originally prescribed for his 15-year-old son's acne. He comes to see his doctor after noticing he has been urinating more frequently than normal ever since taking this drug. Physical examination reveals dry mucous membranes. Blood tests reveal hypokalemia and a pH of 7.2. Urinalysis reveals elevated levels of glucose, protein, calcium, and phosphate. If this condition goes untreated, which of the following is the most likely long-term complication for this patient?

a. Diabetes insipidus  
b. Diabetes mellitus  
c. Osteomalacia  
d. Osteopetrosis  
e. Rickets

correct answer: C

Seeking relief from the flu, this patient self-medicated with tetracycline, which was originally prescribed for his son's acne vulgaris. Expired tetracycline is a known cause of Fanconi's syndrome, a hereditary or acquired disorder of the proximal tubule of the kidney. This leads to defective transport of certain substrates, including glucose, amino acids, sodium, potassium, phosphate, uric acid, and bicarbonate. Characteristic findings include glucosuria, phosphaturia, proteinuria, polyuria, dehydration, hypercalciuria, and hypokalemia. Bicarbonate wasting can lead to renal tubular acidosis, while phosphate wasting can disrupt vitamin D metabolism and bone mineralization. This can result in rickets or impaired growth in children, and osteomalacia, osteoporosis, and pathologic fractures in adults.
A 60-year-old single male who has multiple sexual partners presents complaining of a painless chancre on his penis. His VDRL test returns a positive result. Administration of which of the following is most appropriate?

- a. Amphotericin B
- b. Azithromycin
- c. Erythromycin
- d. Penicillin G
- e. Penicillin V

**Correct answer: D**

Syphilis, in its primary stage, presents with a solitary, painless chancre. It is caused by the spirochete Treponema pallidum. The gold standard for treatment is Penicillin G, which is the parenteral form. Penicillin V is the oral equivalent. Erythromycin and Azithromycin could be used to treat Haemophilus ducreyi, a sexually transmitted pathogen that presents with painful chancroid. Amphotericin B is an antifungal agent.
A 74-year-old man with urinary frequency and urgency has benign prostatic hyperplasia. He refuses operative intervention but agrees to a trial of finasteride therapy. During the trial, synthesis of which of the following substances is most likely to be inhibited?

- a. Androstenedione
- b. Dihydrotestosterone
- c. Estradiol
- d. Estrone
- e. Testosterone

**correct answer:** B

Finasteride inhibits 5-alpha-reductase, interfering with the conversion of testosterone to dihydrotestosterone (DHT) (choice B). DHT is the primary mediator in the pathogenesis of benign prostatic hyperplasia, causing hyperplasia of glandular and stromal cells.

Androstenedione (choice A) is the precursor to both male and female sex hormones. It can be formed from either dehydroepiandrosterone converted by 3-beta-hydroxysteroid dehydrogenase or 17-alpha-hydroxyprogesterone converted by 17,20 lyase.

Estradiol (choice C) is the major estrogen in humans. Estradiol can be formed by the aromatization of testosterone. Additionally, androstenedione can be aromatized to estrone (choice D) and then converted to estradiol by 17-beta-hydroxysteroid reductase.

Testosterone (choice E) is formed by converting androstenedione via oxidoreductase.
A 46-year-old man comes to the physician because of a 2-week history of intermittent dizziness and difficulty standing up. His symptoms began after he started treatment with sildenafil for erectile dysfunction. Physical examination shows no abnormalities. Which of the following is the most likely mechanism of action of sildenafil causing these adverse effects?

a. Decreased basal vascular smooth muscle tone
b. Decreased parasympathetic nerve activity
c. Decreased sympathetic nerve activity
d. Increased basal vascular smooth muscle tone
e. Increased parasympathetic nerve activity

**Correct answer:** A

Sildenafil (Viagra) is a commonly-used medication for erectile dysfunction and increasingly pulmonary hypertension. The drug is a potent cGMP phosphodiesterase 5 (PDE5) inhibitor. Thus, it blocks PDE5, which increases cellular cGMP in the corpus cavernosum. The increased cGMP promotes smooth muscle relaxation and vasodilation, facilitating an erection.

A major side effect of sildenafil is that the above cascade can also occur in the general circulation at the level of the arteriole, thus causing systemic vasodilation. This may result in dizziness and orthostatic hypotension. Such symptoms are especially worsened by nitrates such as nitroglycerin, which is contraindicated in patients taking sildenafil or other PDE5 inhibitors.
A 72-year-old male presents to the physician complaining of waking up multiple times during the night to urinate and having trouble starting his stream. The patient reports that his health food store owner recommended that he try an herbal medication. Which of the following herbal agents did the health food store owner likely recommend to help this patient’s condition?

a. Echinacea  
b. Feverfew  
c. Kava  
d. Saw palmetto  
e. St. John’s wort

**Correct answer:** D

USMLE Step 1 expects a familiarity with herbal agents and complementary and alternative medicine. In this question, the patient's symptoms of urinary frequency and hesitancy are consistent with BPH, benign prostatic hyperplasia. Saw palmetto (choice D) has been reported to help patients with BPH.

The other herbal agents are not recommended for BPH:

Echinacea (choice A) is used for the common cold.

Feverfew (choice B) is used to relieve migraines.

Kava (choice C) is used for chronic anxiety.

St. John’s wort (choice E) is used for depression.
35-years-old female was placed on oral contraceptive, 19-nortestosterone (progestational agent) for the past two years. She will most likely develop?

a. Endometriosis
b. Pelvic inflammatory disease
c. Ovarian cancer
d. Hypertension
e. Endometrial cancer

**Correct answer:** D

D is the correct answer:

Oral contraceptives are known to increase the liver synthesis of angiotensinogen - the most common cause of hypertension in young women. Patients on oral contraceptive are also prone to develop cervical and hepatocellular carcinoma, intrahepatic cholestasis and hepatic adenoma.

A is incorrect: OCPs are protective against endometriosis.
B is incorrect: OCPs are protective against pelvic inflammatory disease.
C is incorrect: OCPs are protective against ovarian cancer.
E is incorrect: OCPs are protective against endometrial cancer.
A 72 year old man with erectile dysfunction is being treated with sildenafil. This drug acts by inhibiting which of the following?

a. 5-alpha reductase  
b. cAMP phosphodiesterase  
c. cGMP phosphodiesterase  
d. testosterone receptors  
e. estrogen receptors

**Correct answer:** C

Sildenafil blocks cGMP phosphodiesterase, which is normally responsible for the breakdown of cGMP. Sildenafil therefore leads to increased levels of cGMP, which have vasodilatory effects to relax smooth muscle. During sexual arousal, increased blood is able to flow through the penis and allow for erection.
A patient with asthma is being treated with a xanthine derivative such as theophylline. The drug relaxes smooth muscle in airways by which of the following mechanisms?

- a. Activating (indirectly) beta2 adrenergic receptors
- b. Blocking sarcoplasmic reticulum calcium release
- c. Inhibiting the ryanodine receptor
- d. Preventing cAMP degradation
- e. Stimulating ATP production

**Correct answer:** D

Theophylline inhibits phosphodiesterase (PDE). PDE hydrolyzes and thus inactivates endogenous cAMP, therefore this is an (indirect) inhibitory effect of the drug. When theophylline blocks PDE, cAMP accumulates intracellularly; calcium uptake is promoted and calcium release from the SR is attenuated by a putative process involving cAMP-induced protein kinase A (PKA) activation.

Xanthine derivatives are also thought to function by increasing histone deacetyases (HDACs) activity. This removes acetylation from pro-inflammatory genes and thus reduces their expression (epigenetically).
A 25 year old woman with a history of asthma complains of worsening asthma attacks. Before, she had symptoms less than twice a week with less than two night attacks a month. These symptoms were easily controlled with her albuterol inhaler. Now, she is symptomatic every day of the week with frequent weekly night attacks. The albuterol inhaler helps control the acute attack, but she is concerned that she needs additional medication. You decide to add low-dose inhaled steroids and a long-acting bronchodilator. Which of the following drugs would be the most appropriate long-acting bronchodilator to add?

- a. Salmeterol
- b. Cromolyn
- c. Theophylline
- d. Zafirlukast
- e. Salmeterol or Theophylline

correct answer: A

Salmeterol (choice A) and Theophylline (choice C) are the only two drugs listed that are considered long-acting bronchodilators. Salmeterol is a long-acting beta-2 agonist that acts as a bronchodilator by increasing cAMP in smooth muscle cells throughout the respiratory system resulting in muscle relaxation. Theophylline is a methylxanthine that also causes smooth muscle relaxation by increasing levels of cAMP. Between the two, salmeterol would be preferred because theophylline has a narrow therapeutic index with risk of cardiotoxicity and neurotoxicity; therefore choice E is incorrect.

Cromolyn (choice B) prevents the release of mediators from mast cells and is therefore not a bronchodilator.

Zafirlukast (choice D) is an antileukotriene that inhibits leukotriene receptors to mediate the inflammatory response and is therefore not a bronchodilator.
You begin to follow a child that was recently diagnosed with cystic fibrosis. You decide to prescribe an inhaled agent to help dissolve the mucus plugging the airways. This agent is also used as an antidote to reverse the toxicity of which of the following drugs?

a. aspirin  
b. acetaminophen  
c. heparin  
d. streptokinase  
e. methamphetamine

**Correct answer:** B  
N-acetylcysteine, or Mucomyst, is a mucolytic agent that works by breaking down disulfide bonds within mucus proteins. It is used in the treatment of patients with conditions involving excessive mucus clogging the airways. For the CF patient above, inhaled Mucomyst is indicated. N-acetylcysteine is also used widely to reverse acetaminophen toxicity. For this purpose, it is typically given intravenously. It helps to restore glutathione reserves to quench the toxic metabolites and therefore protect the liver cells.
A 55 year-old male reports to his primary care physician complaining of a headache, fever and a dry cough for the past 3 days. The patient admits to being a pack a day smoker for the past 35 years, and although he has tried to quit, he has never been successful.

A chest X-ray shows small unilateral infiltrate in the left lower lobe. And blood tests are sent:

- WBC: 14,000
- Hgb: 13g/dl
- Platelets: 400,000
- Na+: 128
- K+: 4.1
- Cl-:100
- HCO3: 23
- BUN:25
- Cr:1.2

What antibiotic would be the most appropriate in this situation?

a. Amoxicillin
b. Azithromycin
c. Ceftriaxone
d. Cephalexin
e. Vancomycin

Correct answer: B

This patient is most likely suffering from Legionnaire's Disease which is an infection with the bacteria Legionella in the spectrum on atypical pneumonias. Legionnaire's is more common in smokers or people who are immune suppressed. Often there is a history of a person being near aerosolized water (air conditioning etc) but it is a bacteria that colonizes in common places, so this history is not necessary. More common in Legionnaire's in comparison to other pneumonias is that the patient often has hyponatremia (as above). Therefore the best treatment for an atypical pneumonia is a macrolide like azithromycin (option B). A fluoroquinolone is also acceptable.

Amoxicillin (A), ceftriaxone (C) and cephalexin (D) are part of the beta-lactam group, but they do not have much activity against atypical pneumonias.

Vancomycin (E) is useful against MRSA and other gram positive bacteria; however, there is no evidence that this patient is suffering from MRSA and therefore, there is no need to use it.
A 3-year-old boy is prescribed a medication by his pediatrician. Five days after beginning this medication, the child develops the rash shown above. The lesions are non-pruritic and non-blistering; at no point does the child develop respiratory distress. Of the following medications, which is most likely responsible for the observed reaction?

- a. Amoxicillin
- b. Cefazolin
- c. Gentamycin
- d. Hydroxychloroquine
- e. Vancomycin

**correct answer:** A

This is the morbilliform (measles-like) “amoxicillin rash.” It typically begins on the trunk >72 hours after beginning the medication, then spreads, and is non-pruritic. As it is not a true allergic reaction, the current regimen may be continued and there is no contraindication to future penicillin use.

Though first-generation cephalosporins like cefazolin (Choice B) may show cross-allergenicity with penicillins, rash is uncommon. Anaphylaxis is a more likely reaction, with onset sooner than four days.

The aminoglycoside gentamycin (Choice C) is classically associated with nephrotoxicity and ototoxicity. Dermatologic adverse events, though rare, are severe and include erythema multiforme, toxic epidermal necrolysis, and Stevens-Johnson syndrome.

Common adverse effects of hydroxychloroquine (Choice D) include GI distress, pruritus, and dizziness. Severe reactions include hemolysis and ocular dysfunction.

Vancomycin (Choice E) is associated with the rash of “red man syndrome,” however, this reaction is infusion-rate dependent. Other adverse effects include chills, fever, ototoxicity and nephrotoxicity.
A 59 year-old man with angina uses sublingual nitroglycerin to relieve his occasional episodes of chest pain. He has never had a myocardial infarction. He tells you that on several occasions his pain has not been completely relieved after taking nitroglycerin, so he has taken double the recommended dose. When he has done this, he has become lightheaded, and most recently, he fainted after standing up. What is the predominant mechanism for his lightheadedness/syncope?

a. Arterial vasoconstriction, increased systemic blood pressure, increased intracranial pressure  
b. Arterial vasodilation, increased afterload, decreased cardiac output, decreased cerebral perfusion  
c. Decreased myocardial contractility, decreased cardiac output, decreased cerebral perfusion  
d. Venous vasoconstriction, decreased preload, decreased cardiac output, decreased cerebral perfusion  
e. Venous vasodilation, decreased preload, decreased cardiac output, decreased cerebral perfusion

**Correct answer:** E

Nitroglycerin (a nitrate) causes relaxation of venous smooth muscle, leading to decreased venous pressures, and decreased venous return. This leads to decreased end diastolic volume (EDV) and less strain on heart muscle, relieving anginal pain. Using too much nitroglycerin could cause venous pooling and an excessive decrease in cardiac output (since VR = CO). The decrease in CO could cause lightheadedness, and when coupled with positional changes, could cause orthostatic syncope.
A healthy 25-year-old man is undergoing an exercise stress test. Which of the following is most likely to occur in this man’s skeletal muscle during exercise?

- a. Decreased capillary hydrostatic pressure
- b. Decreased metabolite concentration
- c. Increased arteriolar diameter
- d. Increased oxygen concentration
- e. Increased vascular resistance

**Correct answer: C**

As skeletal muscle activity increases, the oxygen demand increases. The vascular supply to the area responds by dilating the arterioles in order to bring in more blood and supply the increased oxygen demand.

Choice A is incorrect because hydrostatic pressure in the capillaries is increased as blood flow increases.

Choice B is incorrect because contracting skeletal muscle will produce metabolites and release them into the capillaries.

Choice D is incorrect because contracting muscle cells will take up oxygen from the capillaries.

Choice E is incorrect because the vascular supply to the muscle is increased as the arterioles vasodilate, therefore vascular resistance decreases.
Which of the above figures (A–E) best illustrates the relationship between blood flow and mean arterial pressure (MAP) in a vascular bed that demonstrates autoregulation of blood flow?

a. Figure A
b. Figure B
c. Figure C
d. Figure D
e. Figure E

correct answer: D

Figure D shows a plateau of blood flow as mean arterial pressure (MAP) increases. This illustrates the phenomenon of autoregulation, in which tissues alter pre- or post-capillary resistance to keep blood flow relatively constant in the face of changing MAP. (The curve appears similar to those seen in pH buffer systems.)

Figure A shows a direct relationship between blood flow and MAP. This is an absence of autoregulation.

Figure B shows an inverse relationship between blood flow and MAP. This is abnormal and is not an example of autoregulation.

Figure C shows an increase in blood flow at a constant MAP. This is abnormal and is not an example of autoregulation.

Figure E shows a plateau of blood flow as MAP increases, but high blood flow should be seen with high MAP, not low.
A 45-year-old man has a left ventricular ejection fraction of 25% (N>55%) with diffuse hypokinesis. He has a sedentary life-style. He eats red meat up to 6 times weekly and drinks 4 alcoholic beverages daily. He is 185 cm (6 ft 1 in) tall and weighs 86 kg (190 lb); BMI is 25 kg/m^2. His blood pressure is 90/60 mm Hg. Coronary arteriography shows no evidence of atherosclerosis. To prevent further heart damage, which of the following is the most appropriate recommendation?

a. Aerobic exercise program
b. Avoidance of alcohol
c. Ingestion of more vegetables and decrease in red meat intake
d. Isometric/weight-training exercise program
e. Weight loss

correct answer: B

This patient has heart failure, hypotension, and no signs of atherosclerosis in the setting of alcohol abuse, sedentary lifestyle, and poor dieting. The most likely diagnosis is dilated cardiomyopathy secondary to alcohol abuse. Dilated cardiomyopathy is the most common cardiomyopathy and etiologies include alcohol abuse, thiamine deficiency, coxsackie viral infection, cocaine abuse, doxorubicin toxicity, and idiopathic. Ultimately, patients suffer from systolic dysfunction with eventual cardiac arrest. The most appropriate recommendation for this patient is to remove the source of his cardiomyopathy—alcohol (choice B).

Diet (choice C), exercise (choice A & D), and weight loss (choice E) will definitely have cardio-protective effects; however, removing the etiology of the patient’s heart failure needs to have highest priority in the management of this patient.
A cardiac catheterization is done in a healthy person. The blood sample withdrawn from the catheter shows 60% oxygen saturation, and the pressure recording shows oscillations from a maximum of 26 mm Hg to a minimum of 14 mm Hg. The catheter tip was located in which of the following areas?

a. Ductus arteriosus  
b. Foramen ovale  
c. Left atrium  
d. Pulmonary artery  
e. Right atrium

_correct answer_: D

Blood pressures in the pulmonary artery (choice D) are in the range of 15-30mmHg systolic and 4-12mmHg diastolic, which are pressures that most closely match those found in this patient’s catheterization.

Left atrial (choice C) pressures range from 1-10mmHg, and right atrial (choice E) pressures range from 0-8mmHg. Prior to birth, the ductus arteriosus (choice A) diverts blood from the pulmonary artery to the aorta, and the foramen ovale (choice B) provides a communication between the two atria, equalizing their pressures, and shunting blood from the pulmonary circulation to the systemic circulation.
A 65 year old male with a history of heart failure sees his physician for a routine checkup. He has been on a stable regimen of metoprolol and furosemide to manage his blood pressure and heart failure. A routine ECG in the office shows flattening of the T wave, appearance of a U wave, and ST segment depression. What is the most likely explanation for the ECG findings?

a. High serum magnesium
b. High serum potassium
c. Low serum magnesium
d. Low serum potassium
e. Recent myocardial infarction

**Correct answer: D**

The ECG findings are characteristic of hypokalemia. This man is at risk of hypokalemia because he is on long-term furosemide.

Isolated magnesium abnormalities (A,C) do not typically cause these ECG abnormalities.
(B) High serum potassium typically manifests as peaked T waves, shortened QT interval, disappearance of p waves, and eventually a widened QRS and a sine wave pattern.
(E) Flattening of the T wave and U waves are not typical of MI.
A three-year old male patient who has just been diagnosed with tetralogy of Fallot (TOF) presents to your office for a routine well-child checkup. His condition presented when his parents noticed that he was squatting frequently and seemed to be in distress. After having been given a referral for a pediatric cardiologist, they were informed that the squatting helps to compress the femoral arteries which increases systemic vascular resistance and decreases the right to left shunting present in TOF. When auscultating the heart on physical exam, what is the most likely characteristic of the heart sounds?

- a. Fixed splitting of S1
- b. Fixed splitting of S2
- c. Normal splitting of S2
- d. Paradoxical splitting of S2
- e. Wide splitting of S2

**Correct answer:** E

Wide splitting of S2 (choice E) is most commonly seen in pulmonic stenosis due to lengthening of right ventricular ejection time. This pulmonic stenosis is one of the four components of the Tetralogy. The other four components are ventricular septal defect, overriding aorta, and right ventricle hypertrophy.

Fixed splitting of S1 (choice A) can result from conduction disturbances, hemodynamic, or mechanical causes but is not usually heard on physical exam.

Fixed splitting of S2 (choice B) is usually seen with an atrial septal defect or with right bundle branch block. The mechanisms for each of these is either increased blood flow in the case of an atrial septal defect or a delay in conduction in the case of a right bundle branch block.

In normal splitting of S2 (choice C), you see an increased time between the closure of the aortic valve and the pulmonic valve at the end of systole during inspiration due to pooling of blood in the pulmonic vasculature.

Paradoxical splitting of S2 (choice D) is when the aortic valve closes after the pulmonic valve. This can occur in aortic stenosis due to similar mechanisms described above for the wide splitting found in pulmonic stenosis.
The accompanying image is a synchronous tracing of aortic pressure, left atrial pressure, left ventricular pressure, left ventricular volume, and EKG throughout the cardiac cycle. Which of the following is the best description of events that occur during Time Period 1?

a. Aortic valve closes, mitral valve opens  
b. Aortic valve opens  
c. Diastolic inflow of blood from left atrium to left ventricle  
d. Left atrium contracts  
e. Mitral valve closes

**Correct answer:** E

Mitral valve closes (Choice E) is the correct answer. During Time Period 1, the left ventricular pressure exceeds left atrial pressure, causing mitral valve closure. It is important to note that there is a slight lag between mitral valve closure and aortic valve opening, which occurs when left ventricular pressure exceeds aortic pressure (after Time Period 1 ends).

Aortic valve closes, mitral valve opens (Choice A) is incorrect; these actions occur during Time Period 2, represented by the dichrotic notch of the aortic pressure tracing, and when left ventricular pressure falls below left atrial pressure, respectively. Note that although these actions are associated together, they are not simultaneous.

Aortic valve opens (Choice B) is incorrect; this action occurs after Time Period 1 ends, when left ventricular pressure exceeds aortic pressure. It is important to note that there is a lag between mitral valve closure and aortic valve opening.

Diastolic inflow of blood from left atrium to left ventricle (Choice C) is incorrect; this action occurs between Time Periods 2 & 3, evidenced by the increase in left ventricular volume with relatively constant left ventricular pressure.

Left atrium contracts (Choice D) is incorrect; this action occurs before Time Period 1 (and between Time Periods 3 & 4), causing the observed increase in atrial pressure and ventricular volume.

Adapted from http://commons.wikimedia.org/wiki/File:Cardiac_Cycle_Left_Ventricle.PNG
A 66-year-old man with a history of high cholesterol and high blood pressure presents to the doctor for a routine physical exam. He currently has no health complaints. On physical exam, he is noted to have a systolic murmur heard loudest on the right side of his chest. On further examination, the murmur can also be heard in his neck. The murmur is most likely to be which of the following?

- A. Aortic regurgitation
- B. Aortic stenosis
- C. Mitral regurgitation
- D. Mitral stenosis
- E. Mitral valve prolapse

**Correct answer:** B

The murmur of aortic stenosis is classically described as a harsh, crescendo-decrescendo systolic murmur. It is most audible in the second right intercostal space and radiates to the carotid arteries. Other physical examination signs may include a sustained PMI, precordial thrill, and S4. Patients are often asymptomatic for years, in which case no therapy is required. Once a patient develops symptoms of angina, syncope, or heart failure, surgery is often indicated.

The murmur of aortic regurgitation (Choice A) is classically described as a diastolic decrescendo murmur most audible at the left sternal border. Other physical examination signs include a displaced PMI, water-hammer pulse palpable at the radial and femoral arteries, and S3. Symptoms may include dyspnea on exertion, paroxysmal nocturnal dyspnea, orthopnea, palpitations, and angina.

The murmur of mitral regurgitation (Choice C) is classically described as a holosystolic murmur most audible at the apex of the heart that radiates either to the back or clavicular area. Other physical examination signs may include a laterally displaced PMI, loud P2, and S3 gallop. Symptoms may include dyspnea on exertion, paroxysmal nocturnal dyspnea, orthopnea, palpitations, and pulmonary edema.

The murmur of mitral stenosis (Choice D) is classically described as an opening snap after S2, followed by a low-pitched diastolic rumble and loud S1. With long-standing disease, signs of right ventricular failure can be seen, including JVD, heave, hepatomegaly, ascites, and pulmonary hypertension. Symptoms may include exertional dyspnea, orthopnea, paroxysmal nocturnal dyspnea, palpitations, chest pain, hemoptysis, and thromboembolism.

The murmur of mitral valve prolapse (Choice E) is classically described as a midsystolic or late systolic click followed by a mid-to-late systolic murmur. The murmur is increased by standing and the Valsalva maneuver and decreased by squatting.
A 38-year-old male is brought into the emergency room with mild confusion and disorientation to person, place and time. His diet consists exclusively of salads and coffee. He is found to have an acute dermatitis on his forearms and reports loose, watery diarrhea for the past three days. What vitamin deficiency would you suspect in this patient?

a. Cobalmin  
b. Niacin  
c. Thiamine  
d. Vitamin C  
e. Vitamin D

Correct answer: B

Niacin, a.k.a. Nicotinic Acid or Vitamin B3 produces many biologically active derivatives including NAD, NADP, NAD+ and NADH. Niacin deficiency results in the classic triad of dementia, dermatitis and diarrhea, known as pellagra. Thiamine deficiency (C) results in Wernicke-Korsakoff syndrome often in chronic alcoholics with poor nutrition, while the others present with multiple signs/symptoms.
A 25-year-old woman with previously well controlled type 1 diabetes mellitus develops ketoacidosis 2 days after onset of a urinary tract infection. The most likely cause of the ketoacidosis is decreased efficacy of insulin on muscle, adipose tissue, and liver resulting from antagonism by which of the following hormones?

- a. Androstenedione
- b. Cortisol
- c. Glucagon
- d. Leptin
- e. Thyroxine (T4)

**Correct answer: B**

Acute physical or psychological stress activates the hypothalamic-pituitary-adrenal axis, resulting in increased plasma ACTH and (B) cortisol concentrations. Fever, caused by systemic infection or by pyrogen administration, is a potent stimulus of ACTH and cortisol secretion, while infection causes release of interleukins IL-1 and IL-6, which stimulate hypothalamic CRH secretion, and tumor necrosis factor-alpha, which stimulates ACTH secretion directly.

Cortisol, in turn, acts as a physiological antagonist to insulin by inhibiting glycogenesis, promoting lipolysis, and mobilizing extrahepatic amino acids and ketone bodies. This leads to gluconeogenesis with resultant increased circulating glucose concentrations.
An investigator studying the synthesis of thyroid hormones has developed a transgenic animal deficient in thyroid peroxidase. Within the thyroid gland of this animal, which of the following is most likely to be inhibited?

- a. Cleavage of iodine from iodinated tyrosines in the thyroid cell cytoplasm
- b. Endocytosis of colloid from the follicle lumen into the thyroid cell
- c. Iodide oxidation and binding to tyrosine in the follicle lumen
- d. Movement of iodide from plasma into thyroid cells
- e. Separation of thyroxine (T4) from thyroglobulin in lysosomes

**correct answer:** C

Thyroid peroxidase is the enzyme responsible for iodide oxidation and binding to tyrosine in the follicle lumen.

The other steps in thyroid synthesis including cleavage of iodine from iodinated tyrosines, endocytosis of colloid, movement of iodide from plasma into thyroid cells, and separation of thyroxine from thyroglobulin in lysosomes, do not require thyroid peroxidase and would therefore not be inhibited in the transgenic animal.
A 36-year-old woman dies from an acute adrenal crisis following a surgical procedure. A pituitary tumor was diagnosed 3 weeks ago. At autopsy, the adrenal glands are small, and the cortex is composed primarily of cells from the zona glomerulosa. Which of the following best explains the decreased size of the cortex?

a. Autoimmune destruction
b. Decreased ACTH concentration
c. Decreased cortisol concentration
d. Denervation
e. Granulomatous disease

**Correct answer:** B

(B) In the adrenal gland, the zona glomerulosa produces mineralocorticoids, the zona fasciculata produces glucocorticoids, and the zona reticularis produces androgens. This patient's pituitary tumor was likely causing compression/destruction of ACTH-secreting cells as well as LH/FSH-secreting cells of the pituitary. This caused atrophy of the zona fasciculata and zona reticularis, respectively. The zona glomerulosa, which responds primarily to angiotensin II and serum potassium levels was spared. An acute stressor, such as this woman's surgery, will increase the body's need for cortisol and can lead to acute adrenal crisis in a case such as this.

We are not given any information that would lead us to believe there was autoimmune destruction, denervation, or granulomatous disease. This woman's cortisol deficiency was likely a cause of the tumor in her pituitary rather than a primary adrenal deficiency.
Two chemicals are isolated from the body. When administered at the same time, these chemicals induce gastric acid secretion that is four times as great as that induced by either one alone. The two chemicals are most likely to be which of the following?

- a. Acetylcholine and secretin
- b. Gastrin and cholecystokinin
- c. Gastrin and histamine
- d. Histamine and vasoactive intestinal polypeptide
- e. Secretin and cholecystokinin

**Correct answer:** C

Gastrin and histamine (choice C) both result in increased HCl secretion from parietal cells. Gastrin is produced by the G cells of the antrum and duodenum, and it stimulates parietal cells, as well as increases histamine release from enterochromaffin cells. Parietal cells also have receptors for histamine, which stimulate them to secrete gastric acid.

Acetylcholine will increase gastric acid secretion, but secretin (choice A and E) stimulates secretion of bicarbonate from the pancreas, neutralizing acid.

Cholecystokinin (choice B and E) stimulates gallbladder contraction and pancreatic enzyme secretion.

Vasoactive intestinal polypeptide (choice D) stimulates water and electrolyte secretion from the intestines via cAMP.
After an overnight fast, a child eats a breakfast of cornflakes and milk. The amount of trypsinogen in his duodenum increases because the vagus nerve signals the pancreas to increase which of the following?

- a. Degradation of trypsin
- b. Exocytosis of preexisting trypsinogen
- c. The rate of trypsinogen activation
- d. Transcription of the trypsinogen gene
- e. Translation of trypsinogen mRNA

**Correct answer:** B

During the cephalic and gastric phases that occur after the child eats breakfast, secretions from the pancreas have high concentrations of digestive enzymes in the form of inactive precursors, including trypsinogen. The increased amount of trypsinogen in the duodenum is a result of stimulation of acinar cells by vagal input during cephalic phase and vago-vagal reflex secondary to stomach distention.

Degradation of trypsin does not lead to increased trypsinogen within the duodenum.

The rate of trypsinogen activation does not increase the amount of trypsinogen in the duodenum. It only increases the amount of activated trypsinogen or trypsin in the duodenum. Therefore, the actual amount of trypsinogen in the duodenum decreases as the trypsinogen is activated to trypsin.

Transcription of the trypsinogen gene and translation of the trypsinogen mRNA occur within the acinar cells of the exocrine pancreas and these processes increase the amount of trypsinogen within the acinar cells. However, it is the action of the vagus nerve to increase exocytosis of trypsinogen that leads to an increase of trypsinogen within the duodenum.
A 30 year old male presents with right upper quadrant abdominal pain that worsens after fatty meals. An abdominal ultrasound is done and a diagnosis of cholelithiasis is made. Which cells of the GI tract are responsible for secreting the hormone that stimulates the gallbladder contractions that cause pain in cholelithiasis?

- a. D cells
- b. G cells
- c. I cells
- d. K cells
- e. S cells

correct answer: C

I cells (choice C) of the duodenum and jejunum release cholecystokinin which is a potent stimulator of gallbladder contraction. It also works to stimulate pancreatic sections and decrease gastric emptying.

D cells (choice A) secrete somatostatin and are found in the pancreatic islets and GI mucosa. Somatostatin decreases gastric acid and pepsinogen secretion, decreases pancreatic and small intestine fluid secretion, decreases gallbladder contraction, and decreases insulin and glucagon release.

G cells (choice B) secrete gastrin and are in the antrum of the stomach. Gastrin increases gastric acid secretion, increases growth of gastric mucosa, and increases gastric motility.

K cells (choice D) secrete gastric inhibitory peptide (GIP) and are in the duodenum and jejunum. GIP decreases gastric acid secretion and increases insulin release.

S cells (choice E) secrete secretin and are in the duodenum. Secretin increases pancreatic bicarbonate secretion and decreases gastric acid secretion.
Saliva secretion is stimulated by both sympathetic and parasympathetic innervation. Moreover, the flow rate of saliva secretion is related to the osmolality. If normal salivary osmolality is around 50 mosm/L, what would the osmolality most likely be if there was very little stimulation by the autonomic nervous system?

- a. 25 mosm/L
- b. 50 mosm/L
- c. 60 mosm/L
- d. 75 mosm/L
- e. 100 mosm/L

correct answer: A

If the innervation to salivary glands was decreased, the flow rate would be lower. As the tonicity of saliva decreases with a lower flow rate, the osmolality would be hypotonic (less than 50 mosm/L). The only choice that is less than 50 is choice A. At a higher flow rate, the tonicity of saliva approaches isotonic, but it never becomes hypertonic. Therefore, the other answer choices are unreasonable.
A 6-year-old girl has a history of recurrent pulmonary infections. During physical examination, she coughs frequently, producing thick sputum. Sweat test is positive for increased chloride ion concentration.

For which of the following dietary-related complications is she more at-risk?

a. Angular stomatitis, cheilosis, corneal vascularization
b. Convulsions, peripheral neuropathy
c. Megaloblastic anemia, optic neuropathy, paresthesias
d. Megaloblastic anemia without neurologic symptoms
e. Night blindness, coagulopathy

Correct answer: E

This patient’s positive sweat test indicates she has cystic fibrosis (CF), which often results in malabsorption of fat-soluble vitamins A, D, E, and K. The listed complications result from deficiencies of vitamins A and K, respectively, making “Night blindness, coagulopathy” (choice E) the correct answer.

Angular stomatitis, cheilosis, corneal vascularization (choice A) are complications of vitamin B2 (riboflavin) deficiency. This vitamin is water soluble, and CF is less likely to affect its absorption.

Convulsions, peripheral neuropathy (choice B) are complications of B6 (pyridoxine) deficiency. This vitamin is water soluble, and CF is less likely to affect its absorption.

Megaloblastic anemia, optic neuropathy, paresthesias (choice C) are complications of vitamin B12 (cobalamin) deficiency. This vitamin is water soluble, and CF is less likely to affect its absorption.

Megaloblastic anemia without neurologic symptoms (choice D) is a complication of folate deficiency. This vitamin is water soluble, and CF is less likely to affect its absorption.
A patient diagnosed with Zollinger-Ellison syndrome inquires as to the cause of his disease. As his physician, you explain that which of the following mechanisms is most likely responsible for the disease?

- Destruction of G cells
- Destruction of parietal cells
- Gastric acid-secreting tumor
- Gastrin-secreting tumor
- Somatostatin-secreting tumor

**Correct answer: D**

In Zollinger-Ellison syndrome, a gastrin-producing tumor is responsible for the excess levels of gastrin. Gastrin, a hormone normally secreted by G cells of the stomach and duodenum, stimulates the secretion of gastric acid by the parietal cells of the stomach. Characteristic features of the syndrome include recurrent peptic ulcers, diarrhea, weight loss, and abdominal pain. The majority of gastrin-secreting tumors, or gastrinomas, are found in the "gastrinoma triangle," formed by the cystic duct, junction of the second and third portions of the duodenum, and neck of the pancreas. While 80% of gastrinomas occur sporadically, 20% are associated with MEN I.

Destruction of G cells (Choice A) would lead to a decrease in the production of gastrin. Destruction of parietal cells (Choice B), as occurs in autoimmune gastritis, would result in hypochlorhydria, which, in turn, would cause an elevation in gastrin in an attempt to compensate for the low gastric acid level. Zollinger-Ellison syndrome is the result of a gastrin-secreting tumor, not a gastric acid-secreting tumor. An increase in gastric acid would result in a compensatory decrease in gastrin. Since somatostatin inhibits the release of gastrin, a somatostatin-secreting tumor (Choice E) would result in a decrease in the amount of circulating gastrin.
In the diagram shown, point X indicates the acid-base status of a healthy person. Which of the following is the most likely cause of the condition indicated by point Y?

- a. Adaptation to high altitude
- b. Chronic obstructive pulmonary disease
- c. Diarrhea
- d. Ingestion of a strong acid
- e. Severe prolonged vomiting

**Correct answer: B**

Point Y shows an elevated pCO2, but the pH is slightly below normal, indicating that the patient has compensated and is thus experiencing a chronic acid/base imbalance. In chronic obstructive pulmonary disease (choice B), patients retain CO2 and thus are in a state of chronic respiratory acidosis; the renal compensation is to retain bicarbonate and secrete H+ to correct the pH.

Adaptation to high altitude (choice A) would lead to a higher than normal pH, as a person would try to breathe in more oxygen would hyperventilate, blowing off CO2 and become alkalotic.

In diarrhea (choice C), the patient experiences metabolic acidosis, but the respiratory compensation would lower the pCO2, not raise it.

Similarly, in the case of ingestion of a strong acid (choice D), the patient would hyperventilate to blow off CO2 to compensate for the metabolic acidosis, lowering the pCO2, not raising it.

Severe prolonged vomiting (choice E) leads to loss of acidic gastric contents which would lead to a metabolic alkalosis – since the body never “over-compensated” an acidic pH in the face of metabolic alkalosis is unlikely even with respiratory compensation, and especially not in an acute setting.
An 18-year-old woman has gastroenteritis with nausea and vomiting and is able to ingest only small amounts of water. After 3 days, she develops light-headedness, especially when sitting or standing. Arterial blood gas analysis is most likely to show which of the following sets of values?

- a. pH 7.30, pCO2 28, HCO3 15
- b. pH 7.30, pCO2 55, HCO3 27
- c. pH 7.40, pCO2 40, HCO3 24
- d. pH 7.50, pCO2 30, HCO3 22
- e. pH 7.50, pCO2 47, HCO3 35

**correct answer:** E

Due to the vomiting, this woman is losing HCl (the main stomach acid). Since she is unable to control the vomiting and is not eating she cannot adequately compensate for the acid loss. Therefore, she is in a state of metabolic alkalosis (pH>7.40) which eliminates options A, B, and C. Although her kidneys will eventually compensate by excreting HCO3, this process takes about a week to compensate, and therefore HCO3 will be increased (>24). In order to compensate, the respiratory drive decreases so that the body retains more CO2, and therefore pCO2 is increased (>40). Only option E has increased pH, pCO2 and HCO3.

Option A is metabolic acidosis
Option B is respiratory acidosis
Option C is Normal
Option D is respiratory alkalosis
A 50-year-old woman with a history of ovarian cancer comes to the physician’s office because of swelling in her right leg for the past month. Examination shows edema in the right lower extremity. Which of the following is the most likely cause of the edema?

a. Increased capillary hydrostatic pressure
b. Decreased interstitial hydrostatic pressure
c. Increased capillary oncotic pressure
d. Increased capillary permeability
e. Obstruction of lymph vessels

**Correct answer:** E

In this patient with a history of ovarian cancer, the most likely cause of her edema is (E) obstruction of lymph vessels secondary to her malignancy. While various clinical conditions can cause edema, including congestive heart failure, cirrhosis, and nephrotic syndrome, these disorders are typically associated with pitting edema. This patient presents with nonpitting lymphedema, characterized by its involvement of an isolated limb.

(A) Increased capillary hydrostatic pressure would not cause edema, although decreased capillary hydrostatic pressure would.

(B) Decreased interstitial hydrostatic pressure is unlikely.

(C) Increased capillary oncotic pressure would not cause edema, although decreased capillary oncotic pressure would.

(D) Increased capillary permeability, as in seen in burns, trauma, inflammation, sepsis, and allergic reactions, may cause edema. However, this presentation makes lymphedema much more likely.
A 43-year-old man has a 1-day history of fever and increasing headache, irritability, photophobia, and lethargy. Physical examination shows neck stiffness. Cerebrospinal fluid (CSF) shows 76,000 neutrophils/mm³, occasionally with intracytoplasmic gram-positive cocci. Which of the following substances is most responsible for the CSF neutrophil count?

- a. Bradykinin
- b. C5a
- c. Factor XII (Hageman factor)
- d. Histamine
- e. Leukotriene E4

**Correct answer:** B

C5a is the major factor responsible for neutrophil chemotaxis. Activation of both the classical and alternative complement pathways results in C5a production and eventual membrane attack complex (MAC) formation.
A 26-year-old woman presents at 34 weeks gestational age with dizziness and shortness of breath when lying on her back. In the office, her blood pressure is 124/76 while standing and sitting, but drops to 88/50 when the patient is supine. Which of the following is most likely to explain these findings?

a. Compression of the inferior vena cava by the gravid uterus
b. Dehydration due to hyperemesis gravidum
c. Hemorrhage from placental abruption
d. Hyperventilation due to anxiety about her labor and delivery
e. Pulmonary embolism from hypercoaguable state and venous stasis

**Correct answer:** A

The correct answer is Compression of the IVC by the gravid uterus. This is known as IVC syndrome and is a common cause for decreased supine blood pressure in the third trimester. Women are counseled to sleep on their side to improve symptoms. The other choices can all occur during pregnancy. However, given the decrease in supine blood pressure only, compression of the IVC is the best answer.

Dehydration due to hyperemesis gravidum often resolves prior to the 3rd trimester. If the woman was dehydrated secondary to vomiting, she likely would have decreased blood pressure upon standing and sitting.

Placental abruption often presents with painful contractions and bleeding. The woman would not present with an isolated finding of decreased BP when supine.

Hyperventilation due to anxiety may cause the woman to feel lightheaded and short of breath, but there is no history of anxiety given and this does not explain the decreased BP when supine.

Pulmonary embolism from the hypercoaguable state of pregnancy likely would present with other symptoms of tachycardia and shortness of breath upon standing. While pregnancy is a hypercoaguable state, it is unlikely that a PE would cause isolated decreased BP when supine.
A scientist is investigating transport across cell membranes. She is interested in a type of transport that does not use metabolic energy and does not depend on a sodium gradient, but is carrier-mediated. Which type of transport is she likely studying?

a. Cotransport  
b. Countertransport  
c. Facilitated diffusion  
d. Primary active transport  
e. Simple diffusion

**Correct answer:** C

Facilitated diffusion (Choice C) is a passive process that does not use metabolic energy and does not depend on a sodium gradient. It is, however, carrier mediated.

Cotransport (Choice A) is a secondary active process that uses metabolic energy and depends on a Na gradient (solute moves in the same direction as the sodium across the cell membrane). Countertransport (Choice B) is similar except that solutes and sodium move in the opposite direction.

Primary active transport (Choice D) is not dependent on a sodium gradient but uses metabolic energy.

Simple diffusion (Choice E) does not depend on a sodium gradient or use metabolic energy but is not carrier-mediated.
A healthy 35-year-old woman has a cast removed from her leg after 6 weeks of immobilization. Which of the following best characterizes her gastrocnemius muscle at this time?

- a. Conversion to fast fibers
- b. Decrease in number of fibers
- c. Decrease in number of myofibrils
- d. Increase in mitochondrial content
- e. Increase in number of satellite cells

correct answer: C

Muscle atrophy caused by disuse is usually reversible. It is characterized by a decrease in the density of myofibrils (Answer C) but no change in the overall number of muscle fibers (Answer B). Slow-twitch muscle fibers, fast-twitch fibers and intermediate fibers are differentiated cell types, they do not convert after embryogenesis (Answer A). Long-term exercise causes reactive muscle hypertrophy that may include an increase in mitochondrial content (Answer D). Sarcopenia occurs in old age with failure of satellite cells (Answer E) that help regenerate muscle fibers.
A 4-year-old girl has been unable to eat for 2 days because of a gastrointestinal tract disorder. Which of the following is the major source of fuel being oxidized by her skeletal muscles?

- a. Muscle creatine phosphate
- b. Muscle glycogen
- c. Muscle triglycerides
- d. Serum fatty acids
- e. Serum glucose

**Correct answer:** D

This question tests knowledge of the changes in fuel metabolism that occur with starvation.

Skeletal muscle oxidizes fuels to produce Acetyl CoA, which enters the citric acid cycle to yield ATP for energy during use. We need only to know which fuel provides the Acetyl CoA after 2 days of starvation. Let's go step by step.

In the resting state, skeletal muscles degrade adipose tissue triglycerides (C) and ketone bodies from the liver. During heavy bursts of activity, muscle uses phosphocreatine (A) stores and anaerobic respiration to break down muscle glycogen (B) to lactic acid. Moderately active muscle uses blood glucose (E) in addition to adipose-fatty acids and ketone bodies.

After an overnight fast, glycogen stores in the liver and muscle become depleted. After 24 hours, the blood glucose concentration begins to fall and glucagon is released.

After prolonged starvation (>24 hours), skeletal muscle uses circulating fatty acids (D) from triacylglycerol molecules that were broken down in adipose tissue. This permits the efficient use of fuel for muscle while reserving glucose (and later ketones) for the brain.
A healthy elderly person is most likely to show decreased auditory acuity for which of the following tones?

a. High-frequency tones only
b. Low-frequency tones only
c. Tones of all frequencies
d. Tones presented by air but not by bone conduction
e. Tones presented by bone but not by air conduction

correct answer: A

Presbycusis, or age-related sensorineural hearing loss, affects a majority of the elderly. Hearing loss is usually symmetrical, and is most prominent in high-frequency tones. Diminished perception of any of the other tones in the list suggests a pathological condition and warrants further work-up.
A 55-year-old man develops loss of visual accommodation while taking an antidepressant drug. The most likely cause is blockade of which of the following receptors?

a. Alpha-Adrenergic
b. Beta-Adrenergic
c. Histaminergic
d. Muscarinic
e. Serotoninergic

**correct answer:** D

Visual accommodation occurs by contraction of the ciliary muscle to increase the curvature of the lens. Parasympathetic nerve fibers are responsible for ciliary muscle contraction and these act on muscarinic receptors. The muscarinic receptors are activated by acetylcholine to stimulate contraction of the ciliary body. Many antidepressant drugs have anti-cholinergic properties, thus blocking the muscarinic receptors and resulting in the loss of visual accommodation.

Antidepressant medication also may have anti-histaminergic and anti-serotoninergic properties. However, these two receptors are not found in the ciliary muscle and are not responsible for stimulating visual accommodation.

There are no beta-adrenergic receptors that stimulate the ciliary muscle of the eye. Therefore, blockade of beta-adrenergic receptors would not cause loss of visual accommodation.

There are alpha-adrenergic receptors located in the radial muscle of the eye, but this muscle is not responsible for visual accommodation.
A 33-year-old man has suffered severe head trauma in a motor vehicle accident. Which of the following precautionary treatments would be most appropriate?

a. Hyperventilation  
b. Hypoventilation  
c. Intravenous hypotonic saline  
d. Intravenous thiamine  
e. Lumbar puncture

**Correct answer: A**

In patients with head trauma, you would want to prevent the formation of cerebral edema and increased intracranial pressure. Hyperventilation is often used to produce a respiratory alkalosis, which in turn produces vasoconstriction and reduces the permeability of cerebral vasculature. This helps to mitigate the formation of cerebral edema.

Hypoventilation (B) would produce a respiratory acidosis and result in vasodilation and increased permeability, exacerbating any potential edema.

IV hypotonic saline (C) would likely worsen cerebral edema.

IV thiamine (D) is used to treat Korsakoff’s syndrome, which results from a thiamine deficiency often associated with long-term alcoholism.

Lumbar puncture (E) is contraindicated in patients with head trauma, as a sudden decrease in pressure within the spinal canal may cause uncal herniation.
A 70-kg (154-lb) man on a fixed NaCl intake (200 mmol/day) is given daily injections of a potent mineralocorticoid hormone for 4 days. He has free access to water and consumes his usual caloric intake. Excretion of NaCl is recorded above. Assuming that 1 L of 0.9% saline contains 150 mmol of NaCl and weighs 1 kg, how much will this patient weigh (in kg) at the end of day 4?

a. 66  
b. 68  
c. 70  
d. 72  
e. 74

**Correct answer:** D

Typically, body NaCl level is constant: NaCl excretion equals NaCl intake. However, administration of mineralocorticoid will cause the body to reabsorb more NaCl.

For each day, the difference between NaCl intake and NaCl excretion equals NaCl retention. On day 1 he retains 170 mmol NaCl; on day 2 he retains an additional 110 mmol NaCl; on day 3 he retains an additional 20 mmol NaCl. On day 4, NaCl excretion equals NaCl intake, showing he has reached a new steady-state. Thus, overall he has retained 170 + 110 + 20 = 300 mmol NaCl.

With free access to water and usual caloric intake, the retained NaCl is equivalent to addition of isotonic fluid. Given that 150 mmol NaCl weighs 1 kg, 300 mmol NaCl therefore weighs 2 kg. This 70-kg man will weigh 72 kg (answer D) at the end of day 4.
A 76-year-old man with a history of prostatic hypertrophy has the recent onset of increased difficulty urinating. Symptoms began shortly after he started taking a nasal decongestant orally for cold symptoms. Which of the following types of receptors is most likely to be involved in these adverse effects?

a. alpha1-Adrenergic  
b. beta2-Adrenergic  
c. Ganglionic nicotinic  
d. Nicotinic receptor at the neuromuscular junction  
e. Serotoninergic

**Correct answer:** A

Nasal decongestants are alpha1-adrenergic stimulants that cause smooth muscle contraction, reducing vascular congestion. Benign prostatic hyperplasia is partly due to proliferation of smooth muscle in the bladder neck, urethra, and prostate. Therefore alpha1 blockade is one of the main symptomatic treatments to relieve smooth muscle tension. Main effects of beta2 adrenergic stimulation (Answer B) include bronchodilation and vasodilation. Both sympathetic and parasympathetic ganglionic receptors are nicotinic (Answer C), hence the complex autonomic effects of nicotine. Nicotinic receptor stimulation at the neuromuscular junction (Answer D) causes skeletal muscle contraction, which is not involved in BPH. Serotonergic receptors (Answer E) are not involved in BPH.
A 40-year-old woman receives an intravenous infusion of drug X that selectively constricts the efferent arterioles in her kidneys. Following the infusion, total cardiac output and renal afferent arteriolar tone are unchanged, but renal efferent arteriolar tone and total renal vascular resistance have both increased. Which of the following sets of changes most likely occurred following the infusion of drug X?

a. Decreased GFR, Decreased Filtration Fraction, Same Renal Blood Flow
b. Decreased GFR, Increased Filtration Fraction, Decreased Renal Blood Flow
c. Decreased GFR, Increased Filtration Fraction, Same Renal Blood Flow
d. Increased GFR, Decreased Filtration Fraction, Same Renal Blood Flow
e. Increased GFR, Increased Filtration Fraction, Decreased Renal Blood Flow

correct answer: E

Selective constriction of the efferent arterioles will cause the changes described in choice E. It will increase GFR by causing an increase in the glomerular capillary pressure causing an increase in the net pressure in the glomerulus. There will also be decreased renal blood flow as constriction of either the afferent or efferent arterioles will cause a decrease in perfusion to the kidneys. As the filtration fraction is the GFR divided by renal plasma flow, which is effectively renal blood flow, this number will increase as GFR has increased and renal blood flow has decreased.

The other choices give an incorrect combination of answers.
Serum samples from a healthy woman with a history of regular 28-day menstrual cycles show a peak in the serum concentration of 17beta-estradiol over the past 12 hours. No progesterone is detectable. Within 3 days, which of the following is expected to occur?

a. Cessation of menstruation
b. Decreased basal body temperature
c. Onset of menstruation
d. Ovulation
e. Regression of the corpus luteum

correct answer: D

17 beta-estradiol is the major form of estrogen present in a woman’s body from menarche to menopause. Its role in menstruation is primarily during the proliferative phase (day 1-14) to increase the size of the endometrium and prepare it for the progesterone-dominated secretory phase (day 15-28) during which an embryo will potentially implant and spiral vessels will penetrate the layers of the endometrium. Estradiol peaks on day 13, and a surge of positive feedback on luteinizing hormone (LH) by estradiol during this period causes the follicle to burst on day 14 of the cycle, releasing the egg. Progestone levels are at their lowest during this period, and it is only the secretion of progesterone by the remnants of the follicle, the corpus luteum, that causes a rise in progesterone levels in the second phase of the cycle.
A 25-year-old athlete comes to your clinic and reports that she has not had her period for 8 months. She denies any weight changes and says her period has been normal since it began at age 13. She is 5'6” and weighs 90 pounds. A pregnancy test is negative. What is the most likely cause of her amenorrhea?

a. Early menopause
b. Excessive exercise
c. Hypothyroidism
d. Prolactinoma
e. Resistance to LH and FSH

**Correct answer:** B

While the most common cause of secondary amenorrhea is pregnancy, this patient has had a negative pregnancy test. In athletes, excessive exercise (Answer B) is known to cause amenorrhea. This patient is significantly underweight, which makes this option the most likely answer.

Early menopause (Answer A) is extremely unlikely in a 25-year-old. Hypothyroid patients (Answer C) tend to gain weight. Prolactinomas (Answer D) typically cause physical changes, including headaches and homonymous hemianopia. Resistance to LH and FSH (Answer E) would have manifested as primary amenorrhea during puberty.
A 24-year-old sexually active woman with regular 29-day menstrual cycles presents to the clinic complaining that her period is two weeks late. Physical exam reveals no abnormalities. Pregnancy is confirmed by urine test.

Which of the following mechanisms is most likely responsible for her missed period?

a. Syncytiotrophoblast produces human chorionic gonadotropin (hCG), which stimulates estrogen production by the corpus albicans.

b. Syncytiotrophoblast produces human chorionic gonadotropin (hCG), which stimulates progesterone production by the corpus luteum.

c. Syncytiotrophoblast produces luteinizing hormone (LH), which stimulates estrogen production by the corpus luteum.

d. Zygote produces human chorionic gonadotropin (hCG), which stimulates estrogen production by the corpus albicans.

e. Zygote produces luteinizing hormone (LH), which stimulates progesterone production by the corpus luteum.

**correct answer:** B

The correct answer is B, Syncytiotrophoblast produces human chorionic gonadotropin (hCG), which stimulates progesterone production by the corpus luteum.

Choice A and Choice E are incorrect because the zygote does not produce hCG; further, hCG stimulates the corpus luteum—not corpus albicans. (A corpus albicans is an involuted, non-functional corpus luteum.) Additionally, though hCG stimulates some estrogen production by the corpus luteum, progesterone is the predominant product and is more responsible for absence of menses.

Choice C is incorrect because the syncytiotrophoblast does not produce luteinizing hormone (LH), and LH does not stimulate the corpus luteum. The primary stimulant of the corpus luteum is hCG; though some estrogen is produced, progesterone is the predominant product and is more responsible for absence of menses.

Choice D is incorrect because the zygote does not produce luteinizing hormone (LH), and LH does not stimulate the corpus luteum.
A 59-year-old man with a 40-year history of smoking at least one pack per day presents complaining of progressive shortness of breath over the last few years. On exam, he is found to have lung hyperinflation, prolonged expiration, and hyperresonance to percussion. Which of the following findings is most likely?

- a. Areas of lung collapse
- b. Decreased lung compliance
- c. Decreased residual volume
- d. Loss of small airway patency during expiration
- e. Loss of small airway patency during inspiration

**correct answer:** D

This man likely has emphysema secondary to cigarette smoking. In emphysema, there is destruction of the airspaces distal to the respiratory bronchioles, as well as a loss of elasticity of these airways. As a result of the loss of elasticity, small airways in emphysematous lung regions will collapse due to the increased intrathoracic pressure of expiration. This results in air being trapped distal to collapsed airways. Throughout expiration, these air spaces exert pressure on the chest wall, increasing the anterior-posterior diameter of the chest over time.

Due to air trapping, the tendency of the lungs to collapse is decreased. The exception to this is if a bulla were to rupture, spontaneous pneumothorax could occur, secondarily resulting in atelectasis.

Due to the loss of elasticity and increased lung volume, lung compliance actually increases in emphysema.

Due to air trapping, residual volume is increased.

Small airway patency is maintained during inspiration because there is a negative intrathoracic pressure.
A DIABETIC WOMAN GIVES BIRTH TO A BABY GIRL. WHO IS BROUGHT IN TO EMERGENCY ROOM 3-4 TIMES IN LAST COUPLE OF DAYS, HER BLOOD SUGAR ON ARRIVAL HAD BEEN 36 MG. WHAT IS THE REASON?

a. CONVERSION DISORDER
b. MALINGERING
c. FACTITIOUS DISORDER
d. NEGLECT
e. SOMATIZATION

**Correct answer:** C
A 10 years old diabetic type 1 girl has poorly controlled blood sugar. Her Hb A1c is elevated, despite the fact that she is compliant with her insulin and diet. When you took a detailed history, she accepts trading her lunch with her peers at school. She does not like being treated as a diabetic and thinks that it is not normal or fair. Her maternal aunt who is 36 years old has type 1 DM too. How will you treat her?

a. Ask the school teacher to make sure she finishes her lunch in front of her.

b. Explain to her that with diet and insulin she will be able to live normal life.

c. Increase the dose of her insulin, and tell her that she has to deal with it.

d. Ask her aunt to explain to her that, if she does not comply with her diet and insulin, she will have a lot more medical problems.

e. Arrange for her to attend a summer camp, with children 9-15 years, who have diabetes.

correct answer: E
A 49 year old male with a previous history of paranoid schizophrenia is admitted to the psychiatric ward because of an increase in auditory hallucinations. He has heard voices telling him to set fire to himself. He has been unemployed for five years and has been drinking increased amounts of alcohol since a bitter divorce earlier this year. He has attempted suicide on multiple occasions. His past medical history is significant for poorly controlled diabetes mellitus and hypertension. Which is the best predictor of this patient’s likelihood to commit suicide in the future?

a. Age  
b. Alcohol abuse  
c. Gender  
d. Prior suicide attempt  
e. Recent divorce

**Correct answer:** D  
The strongest indicator that a future suicide attempt is a likely is a history of previous suicide attempts. Male gender is a risk factor for suicide, as well as alcohol, substance abuse, unemployment, recent divorce, and age over 45 years. However, these are less significant than a history of previous suicide attempts.
difficult
   a. yes
   b. yes
   c. yes
   d. yes
   e. yes

*correct answer:* A
yes
A child is observed to walk a distance of 10 feet and climb stairs. Although he cannot yet form full sentences, the child can speak a few words. The age of this child is most likely:

- a. 3 months old
- b. 10 months old
- c. 15 months old
- d. 2 years old
- e. 3 years old

**Correct answer:** C

Walking and climbing stairs are both considered motor milestones for a child that is 15 months of age. Children are typically only able to crawl, and not walk continuously, until at least 1 year of age. At 3 years of age, children should be able to form full sentences and engage in parallel play.
A father brings his 3-year-old daughter to the pediatrician for a well child visit. He is concerned that her level of speech may be delayed for her age. Which of the following would characterize the child's level of verbal and cognitive development as appropriate for a 3-year-old?

- a. Child names body parts
- b. Child uses pronouns
- c. Child speaks in complete sentences
- d. Child strings two words together
- e. Child tells detailed stories

**correct answer:** C

By age 2 children should be able to use pronouns.

By age two years, the child should use about 250 words and should be able to name body parts (choice A). At this age, the child should be able to speak in two-word sentences (ie - "want more") (Choice D).

By age three years, the child should speak in complete sentences (choice C) and use about 900 words.

By age four years, a child should be able to self-express fluently, telling detailed stories (choice E).
A researcher is studying different reactions in patients to psychological stress. Which of the following is an example of an immature ego defense in response to psychological stress?

a. Patient about to undergo surgery jokes about color of hospital gown  
b. Patient recently diagnosed with cancer makes large donation to cancer research  
c. Patient with a desire to cut self and others trains to become a surgeon  
d. Patient with fear of shots does not think about flu shot until arriving at clinic  
e. Patient with newly diagnosed HIV becomes an expert on HIV/AIDS research

**Correct answer:** E

A patient with newly diagnosed HIV who becomes an expert on HIV/AIDS research may be exhibiting intellectualization. This immature defense mechanism allows one to avoid the stressful, emotional aspect of the situation and focus on the intellectual component. For example, a patient who has just been diagnosed with a terminal illness focuses on learning everything about the disease in order to avoid distress and remain distant from the reality of the situation.

A patient about to undergo surgery who jokes about color of hospital gown is using humor as a defense mechanism, one of the mature defenses, in which the subject appreciates the humorous nature of a stressful circumstance. (choice A)

A patient recently diagnosed with cancer who makes large donation to cancer research is exhibiting altruism, one of the mature defense mechanisms in which unpleasant feelings are alleviated by kindness towards others. (choice B)

A patient with fear of shots who does not think about flu shot until arriving at clinic is displaying suppression, a mature defense mechanism in which the subject voluntarily suppresses an idea or feeling from awareness (unlike repression which is involuntary and an immature defense mechanism). (Choice D)

A patient with a desire to cut self and others who trains to become a surgeon is using sublimation, a mature defense mechanism in which unwanted desires are channeled into acts that fit with one's values. (Choice C)
A 16-year-old girl presents to the pediatrician for a routine check-up. She denies feeling depressed but admits that she has been fighting with her mother more often. She reports that she has not menstruated in four months. Physical examination is normal except for excessive growth of downy body hair. Which of the following is likely to be normal in this patient?

- a. Height
- b. CBC
- c. ECG
- d. Lipid panel
- e. X-ray

**correct answer:** A

Anorexia nervosa is an eating disorder characterized by failure to maintain body weight at or above 85% of ideal weight for age and height. Findings indicative of anorexia nervosa include lanugo, or downy body hair, amenorrhea for three or more consecutive months, hypothermia, constipation, and hypotension. Typically, anorexic patients develop their symptoms during their adolescence after the growth spurt, therefore the height (A) of the patient is likely to be normal. CBC (Choice B) in anorexia nervosa may demonstrate anemia and or leukopenia. ECG (Choice C) may show low-voltage T-wave inversion and flattening, ST depression, and prolonged QT intervals. The most common cause of death in anorexia nervosa is cardiac arrhythmias due to hypokalemia. Lipid panel (Choice D) may show hypercholesterolemia and elevated triglycerides. X-ray examination (Choice E) may reveal evidence of early osteoporosis. Anorexia nervosa is also characterized by family conflict, particularly with the mother, normal appetite, increased interest in food, decreased interest in sex, and excessive exercising.
A 77-year-old Caucasian man presents with a chief complaint of fatigue for the last several months. During the course of the interview, his physician discovers that the man was widowed seven months ago and has felt depressed ever since. His daughter and her family live in close proximity and have been trying to persuade him to move in with them. He is hesitant to do so, especially since he has become accustomed to drinking "quite a few" shots of whiskey daily since his wife died. Which of the following factors decreases this man's risk for successful suicide completion?

a. Age  
b. Alcohol consumption  
c. Male sex  
d. Adequate social support  
e. White race

**correct answer:** D

Risk factors for suicide completion include advanced age (Choice A) and being male (Choice C), white (Choice E), and lacking social support. In addition, the use of alcohol or drugs (Choice B) and having a medical illness, organized plan, or prior suicide attempt increase the risk of successful suicide completion. Although women attempt suicide more often, more men die from it.
A female patient presents to your office complaining of decreased energy and inability to fall asleep at night for 3 weeks. She admits to feeling fatigued, which she attributes to her decreased appetite and lack of sleep. In order to consider the DSM-IV criteria for a major depressive episode, which of the following must the patient also display?

a. Feelings of guilt  
b. Inability to concentrate  
c. Loss of interest in most activities  
d. Psychomotor retardation  
e. Suicidal ideation

**Correct answer:** C

Criteria for a major depressive EPISODE is at least 5 of the following 9 symptoms during the same 2 week period, representing a change from previous functioning:

At least 1 of the symptoms MUST be either:
1) Depressed mood OR **2) anhedonia (the loss of Interest or pleasure in most activities).**

the other symptoms include:

3) Sleep disturbance  
4) feelings of Guilt (Choice A)  
5) loss of Energy  
6) inability to Concentrate (Choice B)  
7) change in Appetite  
8) Psychomotor retardation or agitation (Choice D) or  
9) Suicidal ideation (Choice E).

A diagnosis of major depressive DISORDER requires 2 or more such episodes with a symptom-free interval of 2 months.

**HINT:** you can rearrange to capitalized letters of the criteria to the mnemonic "D is SIG E CAPS"
A 15 year old male presents to the physician with severe acne that has not responded to over-the-counter treatments. The patient tearfully tells the doctor that he feels so embarrassed by his skin condition that he has been avoiding leaving the house and skipping school. A former top student, he has begun to fail classes, withdraw from his family and friends, and has been losing weight. Which is the most appropriate response for the physician to give?

a. “It sounds like you are depressed; would you consider seeing a psychiatrist and/or counselor?”
b. “Many people suffer from acne; it’s nothing to be embarrassed about.”
c. “We will treat your acne aggressively and that should help your other problems as well.”
d. “While we will treat your skin condition, why don’t we talk a little bit more about the problems you’re having at school and in your social life”
e. “Your acne really doesn’t look that bad; it probably seems worse to you than it really is.”

**correct answer:** D

This patient is exhibiting signs of depression, including social withdrawal, academic failure, weight loss and sad affect (tearfulness). The physician should certainly treat his acne, but should also address this important psychiatric issue (Choice D).

Choice A acknowledges the mental health issue but does not address the complaint that brought the patient into the office, namely, his acne.

Choices B and E are inappropriate answers because the minimize the patient’s problem and do not legitimize his complaint. Choice E is also particularly condescending, implying that the problem lies in the patient’s mind.

Choice C addresses the acne but does not recognize the severity of the depressive symptoms; this patient should be referred to a mental health specialist immediately for a depression and suicide risk assessment, not wait until his acne resolves in the hopes that his other issues will resolve as well.
Mr A, 38 year old man, previously an occasional drinker has started taking eye opener drinks ever since the death of his wife 2 years ago. He has become less social, and is drunk most of the time. You as a friend are worried about him. What is the most successful way to get him into treatment for his condition?

a. Referal by a doctor  
b. Referal by his son  
c. Referal by employer  
d. No treatment is necessary, normal grief reaction  
e. He is absolutely alrite

**correct answer**: C

The most successful way to get a person into treatment is referral by employer
A 32-year-old man is brought to the emergency department because of multiple nonlethal stab wounds. He is incarcerated and serving a life sentence for murder and armed robbery. After his condition is stabilized, he insists that it is his "right" to remain in the hospital until he is fully "cured." He threatens to harm the attending physician if she endorses his return to prison. Which of the following is the most likely diagnosis?

- a. Adjustment disorder with mixed features
- b. Antisocial personality disorder
- c. Bipolar disorder, manic
- d. Borderline personality disorder
- e. Narcissistic personality disorder

**Correct answer:** B

This patient's history of criminal activity, evidence of recent physical altercation, and acting out with threat of harm toward the attending physician is most suggestive of Antisocial personality disorder. Patients with Antisocial personality disorder demonstrate a pervasive lack of empathy and remorseless disregard toward the rights and well-being of others in their behavior; they often become involved in criminal activity, lie frequently, get into fights, and act in an impulsive and irresponsible fashion.

Patients with Adjustment disorder present with symptoms of anxiety, depression, and/or disturbances of emotion and conduct that manifest within the first 3 months of a specific stressor, and that do not persist beyond 6 months after the resolution of the stressor.

Patients with Bipolar disorder and recent manic episode present with racing thoughts, pressured speech, thoughts of grandiosity often accompanied by increased goal-directed activity, elevated mood that is either euphoric or irritable, and risk-taking behaviors.

Patients with Borderline personality disorder demonstrate a pervasive difficulty with regulating their affect and with forming enduring interpersonal relationships. Their behavior is marked by brief but emotionally intense attachments to others, acts of 'cutting' or other manner of self-harm, and substance abuse. They often report a history of verbal, physical, or sexual abuse or other trauma.

Patients with Narcissistic personality disorder exhibit a pervasive and exaggerated sense of importance and self-worth, one that belies an intense need for admiration and validation from others. They present themselves as flawless and react to criticism with panic and hostility. Like patients with Antisocial PD, patients with Narcissistic PD may show a lack of empathy for other people; however their presentation is characterized more by arrogance and a sense of entitlement than antisocial behavior.
A person attributes his thoughts and emotions to someone else, what is it called?
   a. reaction formation
   b. denial
   c. splitting
   d. interojection
   e. projection

Correct answer: E
projection ;; A person attributes his thoughts and emotions to someone else.
denial; not the reality to penetrate
splitting; good and bad personalities
interojection; identification
reaction formation; unexpected impulse is transferred to opposite.
A 10-year-old boy has been spending much of his time playing with a girl in his class. When his friends tease him about it, he denies having feelings for her, declaring that “Girls are yucky!” Which of the following defense mechanisms is he exhibiting?

- a. Displacement
- b. Dissociation
- c. Reaction formation
- d. Regression
- e. Sublimation

**Correct answer:** C

The correct answer is reaction formation (choice C), an immature defense mechanism in which the unacceptable (showing affection for the opposite gender) is transformed into its opposite.

Displacement (choice A), another immature defense mechanism, involves the subject redirecting an emotion from its original target; e.g. a man kicking his dog because he is mad at his wife.

Dissociation (choice B), another immature defense mechanism, is characterized by separating self from one’s own experiences, often via amnesia or an alternate personality.

Regression (choice D) is an immature defense mechanism in which a person’s actions are unacceptable for his stage of development; e.g. enuresis in a child of this age.

Sublimation (choice E) is a mature defense mechanism in which an unacceptable emotion is expressed in an acceptable outlet. For instance, a man might channel his anger into creating charged artwork.
A 51 year-old man who was previously a successful businessman and devoted husband and father now neglects and abuses his family. At his last visit, he confides to the physician that he abuses alcohol. His wife tells the physician that her husband’s drinking is destroying their family. Of the following responses, which is likely to be the most effective in initiating a discussion about his use of alcohol and its effect on his family?

a. "Did you know most patients who drink as much as you do lose their families?"

b. "Do you feel guilty about what you're doing to your children?"

c. "Do you realize the damage you use of alcohol is doing to your relationships?"

d. "What do you think is the impact of your drinking on your family?"

e. "Your wife says your use of alcohol is destroying your family?"

**correct answer:** D
An 84-year-old woman with Alzheimer's Disease arrives at the hospital accompanied by her daughter. She complains of cough and chest pain. Her temp is 102.4, and she appears to be in mild respiratory distress. On physical examination you notice numerous large contusions on her forearms, trunk, and on the back of her legs. When you ask the patient how she got the bruises, the daughter replies that the patient sometimes “bumps into things.” An chest X-ray reveals lobar consolidation and numerous rib fractures in different stages of healing. Labs are normal with the exception of an elevated white blood cell count. You admit the patient for treatment of pneumonia. Which of the following describes the best course of action?

- a. Advise the daughter to clear the home of any obstacles that the patient may trip over or bump into
- b. Advise the daughter to move the patient to a nursing home where she can be watched more closely
- c. Advise the daughter to encourage bedrest for the patient
- d. Make a note of your findings in the patient’s chart and take no further action
- e. Make a note of your findings in the patient’s chart and report your findings to the authorities

**Correct answer:** E

This is a case of suspected elder abuse. Physicians are required to report suspicion of elder abuse and child abuse to the proper authorities.

Although taking steps to decrease the patient’s fall risk is important, the findings in this patient lead you to suspect elder abuse, so (A), (B), and (C), are incorrect.

Cases of suspected child or elder abuse are exceptions to the patient privilege of confidentiality, so (D) is incorrect. Patient confidentiality may also be breached when you suspect a high likelihood of harm to self or others and in cases of certain communicable diseases.
A 22-year-old college student has had frequent episodes of "sleepiness" over the last 3 months. She frequently falls asleep throughout the day but says that she feels better after each episode. During your conversation with her, she tilts her head and sleeps quietly in the chair for 3 minutes. She has no other complaints. Physical examination is unremarkable. Her BMI is 24. T4 is 8 ug/dL and TSH is 4 ug/dL. Hct is 36%. Which of the following is most likely to be true regarding her condition:

a. This patient’s condition is associated with low frequency, high amplitude waves
b. This patient’s condition is associated with an increased risk of developing right heart failure
c. This patient’s condition is the most common cause of excessive daytime sleepiness
d. This patient’s condition can be treated with CPAP
e. This patient’s condition is associated with low levels of hypocretin (orexin)

**correct answer:** E

The patient described has narcolepsy, which has been associated with low levels of hypocretin. Narcolepsy is characterized by brief episodes of sleep. Patients with narcolepsy often wake up feeling refreshed after each episode.

Narcoleptic sleep attacks begin with REM sleep, which consists of high frequency, low amplitude Beta waves. Low frequency, high amplitude waves describe delta waves so (A) is incorrect.

Answers (B), (C), and (D) all relate to Obstructive Sleep Apnea (OSA). The patient in this case does not demonstrate troubled breathing during sleep, feels better after each sleep episode, does not snore, and is not obese. Thus, OSA is unlikely.

Right-sided heart failure may be a result of Obstructive Sleep Apnea (OSA), so choice (B) is incorrect. Hypoxia in patients with OSA results in pulmonary vasoconstriction and eventual pulmonary hypertension. The pulmonary hypertension can lead to increased work for the right side of the heart, thereby resulting in cor pulmonale.

OSA, not narcolepsy, is the most common cause of excessive daytime sleepiness, so (C) is incorrect.

OSA can be treated with CPAP, so (D) is incorrect.
You went to visit your friend from college who recently had a baby. She named him Johnny. As the mother was gently letting you hold Johnny, he appeared really anxious and started crying once you reached to hold him. Johnny's age is most likely to be:

- a. 2 months
- b. 3 months
- c. 5 months
- d. 7 months
- e. 4 years

**Correct answer:** D

This infant expresses the social milestone of "stranger anxiety", which typically appears around 7-9 months of age. The infant also orients to voice at this age.

A is incorrect. Infants usually have not achieved any social or cognitive milestones by 2 months of age.

B is incorrect. At 3 months, an infant starts expressing the "social smile."

C is incorrect. At 4-5 months an infant starts to recognize people.

E is incorrect. At 4 years of age, a child is typically involved in cooperative play, has imaginary friends, and grooms oneself.
Two young boys are playing at a daycare center. One holds a ball on top of some blocks that the other child has placed on the floor. The second child helps steady the blocks, then the first child lets go of the ball, knocking the blocks down to the floor. They both watch and then repeat the process. These children are most likely

a. 10 months old  
b. 16 months old  
c. 18 months old  
d. 24 months old  
e. 48 months old

correct answer: E
The correct answer is E. The capacity for cooperative play generally does not begin much before the age of 4. Prior to this time (24-30 months), children may play in a parallel fashion, but without real interaction.
A 40 year old Caucasian male with a chronic history of paranoid schizophrenia presents to the mental health clinic. He tells you that his wife stole his bottle of risperidone, which he believes caused him to experience more frequent and intense auditory hallucinations. He says the voices tell him to kill his wife because she cannot be trusted. He admits to having homicidal thoughts about his wife but denies any specific plan for harming her. He requests a refill of his risperidone. What is the most appropriate next step?

a. Admit the patient to the psychiatric ward
b. Call the patient’s wife before filling the risperidone prescription
c. Increase his dose of risperidone
d. Refill his prescription of risperidone and call his wife after he leaves
e. Refill his prescription without calling his wife as he does not have a plan to harm her

**Correct answer:** A

The patient is suffering from chronic paranoid schizophrenia and is therefore prone to hallucinations, delusions, and disorganized thinking. His believes that his wife stole his pill bottle is likely a delusion. What is important is his homicidal ideation, and it is imperative that his wife be protected. Therefore, while he continues to experience command hallucination, he should be admitted to the psychiatric ward to protect his wife. There, he should receive medication adjustment and stabilization.

The patient does need to be restarted on risperidone, but it is more important to admit him to the psychiatric ward. It is also crucial to inform his wife of the potential for harm. Because the patient has not been taking his medication, his current dose of risperidone should be sufficient.

This question is very similar to an USMLE World question. It should be removed.
A 27 year old man with a diagnosis of atypical depression goes to a Mexican restaurant and consumes a meal rich in avocados and cheese. Following the meal he has a few glasses of wine with some friends over dessert. A few hours later he begins to feel uneasy. He starts to feel very nauseated and begins vomiting continuously. At the ED his vitals are BP 200/100; P 120; RR 23; Temp 102. What medication was he likely taking for his atypical depression?

- a. phenelzine
- b. paroxetine
- c. desipramine
- d. amantadine
- e. imipramine

**Correct answer:** A

This patient is obviously suffering from an hypertensive crisis. This is very common in patients taking MAO inhibitors such as phenelzine. When a person on an MAO inhibitor consumes foods rich in tyramine like wines, cheese and avocados; a hypertensive crisis ensues. It is also useful to know that MAO inhibitors are second line agents for atypical depression (depression with weight gain, and excess sleep). Paroxetine is an SSRI and when taken with an MAO inhibitor may result in serotonin syndrome. Amantadine is a parkinson disease medication. Desipramine and imipramine are TCA’s that can also cause a hypertensive crisis when taken with an MAO inhibitor. They are not first or second line for atypical depression.
An 80 year-old male presents with lethargy and anorexia for three days duration. He has a history of atrial fibrillation and has been rate controlled for several years with digoxin. His history is otherwise unremarkable, and his physical exam is within normal limits. After labs are drawn he is found to have a plasma potassium level of 6.0 mEq/L and a digoxin level of 4.0 (normal therapeutic levels are between 0.8 - 2.0).

In the setting of hyperkalemia, which of the following is true?

- a. Plasma digoxin levels will be higher than expected because K+ and digoxin compete for the same target, the membrane-bound calcium channel
- b. Plasma digoxin levels will be higher than expected because K+ and digoxin compete for the same target, the Na+/K+ ATPase
- c. Plasma sodium levels will fall because digoxin overdose is associated with Na+ channel depolarization and opening
- d. Plasma sodium levels will rise because digoxin overdose is associated with increased expression of the Na+/K+ ATPase protein
- e. Plasma potassium levels have no effect on plasma digoxin levels

**Correct answer:** B

The correct answer is B. Digoxin acts by binding to the Na+/K+ ATPase protein, which in turn increases intracellular concentrations of Na+. This decreases the activity of the Ca++/Na+ exchanger and leads to increased intracellular Ca++ levels and an increased inotropic effect in the myocardium. When hyperkalemia is present, such as in the setting of acute renal failure here, K+ competes with digoxin for its binding site (extracellularly) and increases the plasma levels of digoxin, since less digoxin is bound to its target.

Choice A is incorrect; digoxin is not selective for any membrane-bound calcium channel

Choice C is incorrect; digoxin does not bind or depolarize the membrane sodium channel

Choice D is incorrect; Digoxin does not regulate or enhance gene or protein expression

Choice E is incorrect; Potassium levels do have an effect on plasma digoxin levels; see above explanation.
basic science: Biochemistry  

**question:** 41

clinical science: Endocrine

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aaaaa

a. a
b. b
c. a
d. c
e. d

**correct answer:** A

aa
A 14-year-old boy is brought to the emergency room by his parents. They found him passed out on the floor when coming home from work. They inform you that their son has type 1 diabetes. They also say he has been having a hard time coping with having a chronic disease and has stopped taking his insulin. Which of the following will take up glucose through a high-affinity, insulin-independent transporter?

- a. Adipose tissue
- b. Kidney
- c. Liver
- d. Red blood cells
- e. Skeletal muscle

**Correct answer:** D

Red blood cells (choice D) take up glucose independent of insulin levels with a high affinity transporter on their cell membrane. The transporter involved is the GLUT-1 transporter and is also found in the endothelial cells of the blood brain barrier.

Adipose tissue (choice A) and skeletal muscle (choice E) both use the GLUT-4 transporter which is insulin dependent. In the presence of insulin, the GLUT-4 transporter is released from intracellular storage sites to the cell membrane to facilitate passive diffusion of glucose into the cell.

Kidney cells (choice B) and liver cells (choice C) both contain GLUT-2 transporters. Although this is also an insulin-independent transporter, it is a high capacity, low affinity transporter.
THYROID HORMONES ARE DERIVED FROM WHICH OF THE FOLLOWING AMINO ACIDS?

a. THYROSINE  
b. CYTOSINE  
c. IODINE  
d. TYROSINE  
e. THYMINE

**correct answer:** D  
TYROSINE IS THE ONLY AMINO ACID WHICH IS PRECURSOR TO THYROID HORMONES, THE REST OF ANSWERS ARE NOT AMINO ACIDS.
A 45-year-old woman of normal habitus with no prior medical history during her schedule OB/GYN exam mentions to her doctor that she has not had her period recently although she has been abstinent. She has also noticed increased facial hair and oily skin.

Concerned that she might be going through menopause, she mentions that she has taken a herbal preparation of black cohosh that her friend gave to her two days ago. Upon searching the medical literature you discover that the active ingredient in black cohosh belongs to the saponin group, a group of amphipathic glycosides with properties similar to flutamide and surfactant. Assuming that this preparation has therapeutic efficacy, what is the mechanism by which the patient’s self-medication could augment the indicated medication for the condition defined by her presumptive diagnosis?

- Suppression of insulin release
- Blocking effects of androgens
- Increase in progestins
- Decrease in GnRH pulses
- Increase in FSH levels

correct answer: B

The patient is presenting with symptoms suggestive of PCOS, caused by an excessive LH/FSH ratio. Most treatments, herbal or not, for menopause aim to supplement and regulate the bodies estrogen and progesterone levels. The question stem indicates that the herbal remedy functions like flutamide, an anti-androgen. The observation that black cohosh also has similar properties to surfactant may have pharmacological ramifications but functions here as a distraction. The patient is exhibiting signs of androgen excess (hirsutism, acne) that would normally be treated by anti-androgens. Although she did not take black cohosh for this purpose, and should be counseled as to its proper use, the herbal remedy has a similar enough efficacy and mechanism of action to the indicated therapy that it would be expected that this herbal remedy could help given a controlled administration of longer than two days.

For answer choice A: The patient is of normal habitus and has no history of insulin resistance. Furthermore, her main complaints are androgenic symptoms not insulin-related syndromes.

For answer choice C: It is possible that this therapy could cause an increase in progestins, but the only research the doctor found related black cohosh to estrogen levels.

For answer choice D: There is no data to support an inference about the frequency of decreased GnRH pulses. Although lower frequencies favors FSH (implying less LH release) this is a very subtle mechanisms not directly mentioned in the question stem.

For answer choice E: Like answer choice D, this is a possible mechanism of action but there is not data in the question stem to support this.
A 29 year old female with type 1 diabetes goes on a trip to the Caribbean and loses her insulin on the way. Three days after her last insulin dose she goes to the hospital. Lab work shows hyperglycemia and increased anion gap metabolic acidosis. Which of the following pathways is mainly responsible for providing the substrate of her acid-base disorder?

a. B-oxidation of fatty acids
b. Catabolism of branched chain amino acids
c. Citric acid cycle
d. Gluconeogenesis
e. Glycogenolysis

**Correct answer:** A

This patient is in diabetic ketoacidosis which is secondary to an increase in acetoacetate and B-hydroxybutyrate, the anions responsible for the increased anion gap. Ketogenesis occurs in the liver and requires acetyl CoA derived from the B-oxidation of fatty acids.

None of the other pathways produce organic acids that are released into the blood. Look up the balanced equations (input-output) for each pathway or cycle.

Muscle is the primary source that catabolizes branched chain amino acids (answer B) but all the products released into the blood are amino acids, not ketone bodies. An increase in branch chain keto acid derivatives occurs in maple syrup disease.

Each turn of the citric acid cycle (answer C) produces two carbon dioxide molecules, which are released into the blood but do not contribute to an anion gap. Other cofactors produced by the cycle (GTP, NADH, FADH2, etc) stay in the cell.

Gluconeogenesis (choice D) is incorrect. This pathway is partially responsible for hyperglycemia in diabetic ketoacidosis, but the only product released into the blood is glucose.

Glycogenolysis (choice E) is incorrect. It is an initial cause of hyperglycemia but plays no role in ketogenesis. The only product released into the blood is glucose.
Following a fad diet meal of skim milk, an adult female patient experiences abdominal distention, nausea, cramping, and pain followed by a watery diarrhea. What condition is most likely present?

- a. Lactase deficiency
- b. Lipoprotein lipase deficiency
- c. Maltose deficiency
- d. Sialidase deficiency
- e. Steatorrhea

**Correct answer:** A

In many populations, a majority of adults are deficient in lactase (A) and hence intolerant to the lactose in milk. With no transporter existing for lactose in its disaccharide form, and no lactase enzyme to break it down into galactose and glucose, lactose accumulates in the small intestine. This creates an osmotic effect, leading to an outflow of water into the gut. This causes the clinical symptoms of bloating and diarrhea.

Steatorrhea (E), or fatty stools, is caused by unabsorbed fat, which can occur following a fatty meal in persons with a deficiency of pancreatic lipase. Lipoprotein lipase deficiency leads to hypertriglyceridemia. Sialidase deficiency (D) causes accumulation of sialic acid–containing proteoglycans and neurodegeneration.
A 6-year-old boy had meconium ileus at birth. He has now developed chronic productive cough and bulky, fat-containing stools. Which of the following is he most at risk for?

- a. Beriberi
- b. Pellagra
- c. Rickets
- d. Scurvy
- e. Vitamin B12 deficiency

**Correct answer:** C

The most common cause of meconium ileus is cystic fibrosis. The abnormal CFTR channel in cystic fibrosis causes a pancreatic enzyme deficiency that prevents fat digestion and leads to fat excretion, or steatorrhea. In addition to fats themselves, the fat-soluble vitamins, K, A, D, and E, cannot be absorbed. Developing children are at risk for Vitamin D deficiency, or rickets (Answer C). The rest of the choices are water-soluble vitamins. Malabsorption syndromes affect these vitamins as well, although not as severely.

Beriberi, aka B1 or thiamine deficiency, is often associated with alcoholism.

Pellagra, aka Vitamin B3 or niacin deficiency causes the 3 Ds: diarrhea, dermatitis, and dementia.

Scurvy aka Vitamin C or ascorbic acid deficiency, develops in fruit and vegetable deficient diets.

Vitamin B12 deficiency causes anemia and neurologic symptoms, and can be due to autoimmune attack of parietal cells of the stomach (which secrete intrinsic factor, which is necessary for B12 absorption), surgical resection of the ileum (where B12 absorption takes place), or inadequate B12 intake.
A 12-month old child presents to clinic with vomiting, hepatomegaly, hypoglycemia and jaundice. The parents state that they did not notice a significant change in their child’s health until about the time they started feeding him fruit juice. What enzyme deficiency is this child presenting with?

a. Aldolase B  
b. Fructokinase  
c. Galactose-1-Phosphate Uridyl Transferase  
d. Glucose-6-Phosphate Dehydrogenase  
e. Hexokinase

**Correct answer:** A

This child is presenting with hereditary fructose intolerance resulting from a deficiency in Aldolase B. This enzyme is necessary to convert fructose-1-Phosphate to Dihydroxyacetone-Phosphate and Glyceraldehyde. In its absence fructose is still being taken into cells and phosphorylated by fructokinase. The Fructose-1-Phosphate builds up in the cells of the liver. Treatment is simply the avoidance of food/drink that contains fructose.

Note that a deficiency in Fructokinase is benign as fructose cannot be trapped within the cells and is excreted via the urine (fructosuria).
A 12-month old boy presents with vomiting, jaundice, hepatomegaly, failure to thrive, and vision impairment. The attending physician worries about the possibility of mental retardation in this child. The parents state that they did not notice a change in their child until they recently switched his diet from formula to whole milk. The physician suspects the child may have a deficiency in which enzyme:

- a. Aldolase B
- b. Fructokinase
- c. Galactose-1-Phosphate Uridyl Transferase
- d. Glucose-6-Phosphate Dehydrogenase
- e. Pyruvate Kinase

**correct answer:** C

This child is presenting with GALT deficiency. This is the most common cause of galactosemia. Treatment consists of the complete removal of galactose from the diet (i.e. lactose in milk).

The vision problems result from buildup of galactitol in the lenses of the eye (forming cataracts). Galactitol is formed by aldose reductase in order to reduce the levels of galactose in cells.
A 3-day old male infant in the NICU is unresponsive, febrile, and tachypnic. The infant begins seizing every few hours. The attending physician orders several tests of the blood and urine which reveal increased orotic acid and decreased BUN (blood urea nitrogen). This infant is most likely deficient in which of the following enzymes:

- a. Aldolase B
- b. Galactokinase
- c. Glucose-6-Phosphate Dehydrogenase
- d. Ornithine Transcarbamoylase
- e. Phenylalanine Hydroxylase

**Correct answer:** D

This infant is presenting with Ornithine Transcarbamoylase (OTC) deficiency. Without this enzyme the body cannot eliminate ammonia and produces excess orotic acid in order to deplete the large amounts of carbamoyl phosphate that build up from the defunct urea cycle.

This is an X-linked recessive disorder and typically presents within the first few days of life.
The human body has 2 enzymes capable of phosphorylating glucose (the first step of glycolysis), hexokinase and glucokinase. Glucokinase is only found in hepatocytes and the beta-cells of the pancreas. Glucokinase differs from hexokinase in that it has:

- High affinity for glucose (high Km); High velocity (High Vmax)
- Low affinity for glucose (Low Km); High velocity (High Vmax)
- Low affinity for glucose (high Km); High velocity (High Vmax)
- Low affinity for glucose (high Km); Low velocity (Low Vmax)

**Correct answer:** D

This is important for the proper function of the liver and pancreas. Glucokinase’s low affinity and high max velocity make it a great sensor of blood glucose levels. The pancreas and liver need to have this ability in order to control the rates of glycogenolysis, gluconeogenesis, and insulin production.
A patient presents to clinic with muscle cramping, hypoglycemia, and vomiting. A liver biopsy reveals large deposits of triacylglycerols. This patient likely has a deficiency in which of the following enzymes:

- Acyl-CoA Synthetase
- Carnitine
- Citrate Synthase
- Glycogen Phosphorylase
- Lipase

**Correct answer:** B

This is the common presentation of a carnitine deficiency, an autosomal recessive disorder. Without carnitine the mitochondria within hepatocytes cannot transport fatty acids into the mitochondrial matrix to undergo beta-oxidation; hence they triacylglycerols backup and accumulate in the liver.
An African American medical student goes to Central America on a medical mission trip. There, he develops weakness and dark urine soon after taking primaquine for antimalarial prophylaxis. This hemolytic reaction is likely due to:

a. Concomitant scurvy  
b. Diabetes  
c. Glucose-6-phosphate dehydrogenase deficiency  
d. Glycogen phosphorylase deficiency  
e. Vitamin C deficiency

Correct answer: C

Glucose-6-phosphate-dehydrogenase deficiency (Answer C) is the most common enzyme deficiency in the world. This erythrocyte deficiency is most prevalent among African and Mediterranean males. Without glucose-6-phosphate dehydrogenase, the pentose phosphate pathway is unable to produce NADPH. Without the reducing capability of NADPH, erythrocytes have no protection from oxidizing agents, thus leading to hemolysis. This X-linked recessive deficiency is often diagnosed when patients develop hemolytic anemia after receiving oxidizing drugs such as primaquine or after eating oxidizing substances such as fava beans.
A 50 year old woman presents with a history of chronic bloody diarrhea. She denies massive amount of blood per rectum. Physical examination of the heart reveal a murmur that starts immediately after the first heart sound, gradually rises in pitch and gradually decreases before the second heart sound. She denies shortness of breath and abdominal pain. This combination of findings is most consistent with which of the following?

a. Colon carcinoma  
b. Congestive heart failure  
c. Coagulation abnormality  
d. Rheumatic disease  
e. Ischemic colitis

**Correct answer:** C

This patient has Heyde's syndrome, a condition involving aortic valve stenosis associated with gastrointestinal bleeding from colonic angiodysplasia. The latter causes painless bleeding that may manifest as melena (brownish, tarry, foul smelling stools), hematemesis (vomiting blood), or hematochezia (fresh blood in stools). Von Willebrand factor is a protein involved in blood coagulation, as it binds factor VIII. In Heyde's syndrome, von Willebrand factor (vWF) is proteolysed due to high shear stress in the highly turbulent blood flow around the aortic valve. Angiodysplasia is a vascular anomaly characterized by dilated, tortuous submucosal blood vessels lined with endothelial cells, but no smooth muscle cells. Because the bleeding tend to occur in the veins, it is usually not massive, which would be the case for arterial bleed. Colonic angiodysplasia most often originate in the ascending colon or cecum. vWF is most active in vascular beds with high shear stress, such as angiodysplasias, and deficiency of vWF increases the bleeding risk from such lesions. In mild cases, patients are treated with desmopressin, which promotes the release of vWF.

A is incorrect; although colon cancer could cause bloody stools, it does not explain the heart murmur.

B is incorrect; congestive heart failure could develop secondary to aortic stenosis, but does not explain the bloody stools.

D is incorrect; rheumatic disease most often leads to mitral valve abnormalities, which is not suggested in this case. Also rheumatic fever does not explain the gastrointestinal findings.

E is incorrect; a cardinal finding of ischemic colitis is abdominal pain, which is not a complaints in this case.
The substitution of valine for glutamate at position 6 on the two α-chains in sickle cell hemoglobin causes which of the following?

a. Decreased polymerization of deoxyhemoglobin
b. Increased electrophoretic mobility at pH 7
c. Increased solubility of deoxyhemoglobin
d. More flexible red blood cells
e. Unchanged primary structure

**Correct answer:** B

The carboxyl of glutamate at position 6 on the α-chain of normal hemoglobin is dissociated and negatively charged at pH 7. Substitution of uncharged valine for glutamate by mutation produces sickle cell hemoglobin, which is less negatively charged and has an increased electrophoretic mobility. Polymerization of the deoxygenated form of sickle hemoglobin occurs owing to the alteration of primary structure caused by the valine substitution. The insoluble, polymerized hemoglobin causes the erythrocyte to lose flexibility and to become rigid and sickle-shaped. The brittle cells produce anemia and block capillaries.
If all SS individuals in the Minnesota population were sterilized, the SS genotype frequency in the next generation would be which of the following?

- a. Reduced by $\frac{2}{3}$; Reduced
- b. Reduced by $\frac{1}{2}$; Reduced
- c. Reduced by $\frac{1}{3}$
- d. Reduced to 0
- e. Approximately the same

**Correct answer:** E

Even if SS individuals were prevented from contributing to the next generation by sterilization, breeding between AS individuals would replenish SS genotype frequencies. This stability of populations in accord with the Hardy-Weinberg law is often referred to as the Hardy-Weinberg equilibrium. During the decades of 1900 to 1920 in America, the eugenics movement succeeded in passing laws obligating sterilization of those with mental disabilities. These laws were based on two false premises—the idea that mental retardation is always due to Mendelian transmission (ignoring chromosomal and multifactorial disease) and the idea that elimination of affected people will always change gene frequencies.
A newborn infant presents with poor feeding, vomiting, jaundice, and an enlarged liver. The urine tests positive for reducing substances, indicating the presence of sugars with aldehyde groups. Which of the following processes is most likely to be abnormal?

a. Conversion of glucose to galactose
b. Conversion of lactose to galactose
c. Excretion of glucose by the kidney
d. Excretion of galactose by the kidney
e. Conversion of activated galactose to activated glucose

**Correct answer:** E

This infant may have galactosemia (230400), a deficiency of galactose-1-phosphate uridyl transferase (GALT). Galactose from lactose in breast milk or infant formula is phosphorylated by galactokinase, activated to uridine diphosphogalactose (UDP-galactose) by GALT, and converted to UDP-glucose by UDP-galactose epimerase. The elevation of galactose metabolites is thought to cause liver toxicity, and their urinary excretion produces reducing substances. Infants with the signs and symptoms listed are placed on lactose-free formulas until enzyme testing is complete. Deficiencies of epimerase or kinase can cause mild forms of galactosemia.
A previously healthy African-American soldier experiences sudden back pain and pink urine 2-3 days after taking dapsone for leishmaniasis. A Coombs test is negative. The deficient enzyme is the rate-limiting step of what metabolic pathway?

a. Cori cycle
b. Gluconeogenesis
c. Glycogenolysis
d. Glycolysis
e. Pentose phosphate pathway

**Correct answer:** E

This soldier has G6PD deficiency, an X-linked recessive condition that makes red blood cells susceptible to oxidative damage. Cardinal signs are Heinz bodies and bite cells. Glucose-6-phosphate dehydrogenase is the rate-limiting step of E) the pentose phosphate pathway, also known as the hexose monophosphate shunt.

A) The Cori cycle transforms the lactate made during anaerobic glycolysis into glucose. The rate-limiting step of the Cori cycle, which happens in the liver, is lactate dehydrogenase. Deficiency of this enzyme causes autosomal recessive exercise intolerance, myoglobinuria, and a skin rash.

B) The rate-limiting step of gluconeogenesis is fructose-1,6-bisphosphatase. Autosomal recessive deficiency of this enzyme causes fasting hypoglycemia, metabolic acidosis (ketosis), and fatty liver. Individuals with this condition cannot utilize fructose or 3-carbon chains like glycerol for fuel.

C) The rate-limiting step of glycolysis (aka the Embden-Meyerhof pathway) is phosphofructokinase-1. This enzyme is deficient in glycogen storage disease type VII (Tarui’s disease), an autosomal recessive condition causing exercise intolerance, hemolytic anemia, and hyperuricemia. It is the rarest GSD.

The most common enzyme deficiency in glycolysis is of pyruvate kinase. This autosomal recessive condition causes extravascular hemolysis, hemolytic anemia, and jaundice from birth.

D) The rate-limiting step of glycogenolysis is glycogen phosphorylase. Autosomal recessive deficiency of this enzyme causes glycogen storage disease type IV (McArdle’s disease), with exercise intolerance.

In contrast, the rate-limiting step of glycogen synthesis is glycogen synthase. Autosomal recessive deficiency in this enzyme causes glycogen storage disease type 0. Unlike the other GSDs, however, this condition does NOT cause build up of glycogen!

Finally, the most common GSD is type I (von Gierke’s disease), due to autosomal recessive deficiency in glucose-6-phosphatase. It causes hypoglycemia, liver and kidney enlargement, and lactic acidosis.
A 4-year old African-American male presents to clinic with jaundice. A CBC shows decreased red blood cell count with Heinz bodies present within the cells on a blood film. The physician suspects an X-linked enzyme deficiency underlying these symptoms given that they first became present following the recent treatment with sulfadiazene for a UTI. What enzyme is this patient deficient in?

a. Aldolase B  
b. Galactokinase  
c. Galactose-1-Phosphate Uridyl Transferase  
d. Glucose-6-Phosphate Dehydrogenase  
e. Pyruvate Kinase

**Correct answer:** D  
The patient is presenting with symptoms of hemolytic anemia following the recent treatment with a sulfonamide. G6PD deficiency is X-linked recessive and is more prevalent in African-American Populations. This is the most common glycolytic deficiency (accounting for nearly 90% of all cases). G6PD is necessary to send glucose through the Pentose Phosphate Pathway (HMP Shunt). A deficiency results in decreased NADPH levels, which is necessary to keep glutathione (GSH) reduced. Without regenerated GSH the RBC’s become more susceptible to oxidative stress.
Basic science: Biochemistry

**Question:** 1005

Clinical science: Hematology

**Kjblk**

a. q
b. g
c. k
d. o
e. p

**Correct answer:** D

ibuitu    rhgf ikbi
A 6 month-old girl is brought in by his mother who has noted a protruding abdomen, irritability, and recent inability to lift her head unassisted. Examination reveals a healthy appearing girl with a distended abdomen and cervical lymphadenopathy. Funduscopy reveals a cherry red macula. Which of the following is the best diagnosis?

- a. Glucocerebrosidase deficiency
- b. Hexosaminidase A deficiency
- c. Iduronate-2-sulfatase deficiency
- d. Iduronidase deficiency
- e. Sphingomyelinase deficiency

correct answer: E

This child is presenting with Neimann-Pick's disease, characterized by irritability, regression of developmental milestones, hepatosplenomegaly, lymphadenopathy, and a cherry red macula.

Glucocerebrosidase deficiency (choice A) causes Gaucher's disease, which is not associated with a cherry red macula.

Hexosaminidase A deficiency (choice B) is responsible for Tay-Sachs disease, which is similar to Neimann-Pick's disease down to the cherry red macula. However, Tay-Sachs disease does not feature hepatosplenomegaly or lymphadenopathy.

Iduronate-2-sulfatase (choice C) and iduronidase (choice D) deficiencies cause Hunter's and Hurler's syndromes, respectively, and are characterized by facial coarsening. Hunter's syndrome commonly involves corneal clouding. Neither involve a cherry red macula.
A reaction between substrate and enzyme follows Michaelis-Menten kinetics. A competitive inhibitor of the enzyme is introduced into the reaction. What is the likely result?

a. Decreased Vmax
b. Increased Michaelis-Menten constant (Km)
c. Increased Vmax
d. Increased reaction velocity
e. No change

correct answer: B

The M-M equation is derived from this reaction scheme:

\[ E + S \leftrightarrow ES \rightarrow E + P \]

As more S is added, the first step is pushed to right until, at very high S the enzyme is "saturated," so that it is all in the ES complex. Under these conditions the velocity is maximal--Vmax is reached. Vmax is defined as k(cat)E(total), where k(cat) is the intrinsic rate constant for the second step. Km is most easily thought of as the concentration of S that produces 0.5 Vmax. (Find the point on the curve where v= 0.5 Vmax and drop a vertical line to the S axis. The value of S = Km).

A competitive inhibitor competes for the substrate site. The reaction scheme is:

\[ EI \leftrightarrow I + E + S \leftrightarrow ES \rightarrow E + P \]

Increasing I pushes E into the EI form; increasing S pushes E into the ES form. At sufficiently high S, all the enzyme can be pushed into the ES form so that Vmax can still be seen. But if I is present, more S will be required to get there. If more S is required to saturate the enzyme, more S is required to half-saturate the enzyme. By definition, Km appears to go up. (The new Km is called a "Km-apparent.")
The figure above illustrates a G-protein coupled receptor embedded within a lipid membrane. Where would the largest cluster of nonpolar amino acids be found?

- Amino terminus (NH₃⁺)
- Carboxy terminus (COO⁻)
- Cytoplasmic Loop
- Extracellular Loop
- Transmembrane region (1, 2, ..., 7)

**Correct answer:** E

The lipid bilayer is strongly hydrophobic; therefore, non-polar amino acids would cluster here. Polar amino acids prefer binding to water and other polar molecules. A, B, C, and D are all regions with easy access to water.
A child presents with low blood glucose, enlarged liver, and excess fat deposition in the cheeks. A liver biopsy reveals excess glycogen in hepatocytes. Deficiency of which of the following enzymes might explain this phenotype?

- a. alpha-1,1-galactosidase
- b. alpha-1,1-glucosidase
- c. alpha-1,4-galactosidase
- d. alpha-1,4-glucosidase
- e. alpha-1,6-galactosidase

correct answer: D

This question is flawed. Although choice D does identify a glycogen storage disease--Type II, or Pompe's-- the signs and symptoms are not a good fit. Pompe's is unusual in that it is not a defect in the major pathways of glycogen synthesis and degradation, but a defect in the minor pathway of lysosomal degradation. Hypoglycemia is usually not seen because the normal pathway of glycogenolysis is working. Though we usually associate glycogen stores with muscle and liver, all tissues have some, and in this disease massive amounts slowly accumulate in all tissues. Accumulation in the heart is most damaging.
A pair of new parents bring in their one-month-old son saying he has been acting strangely since being brought home from the hospital. They report that the child has been not eating well and seems lethargic. The physical exam is normal except for mild hypotonia. Blood tests taken the week before show a significantly elevated lactate level. Due to a high suspicion for pyruvate dehydrogenase deficiency, a functional enzyme assay is ordered. The diagnosis is confirmed. Which one of these amino acids will the doctor recommend increasing in the child’s diet?

- Alanine
- Glutamate
- Histamine
- Lysine
- Tryptophan

correct answer: D

In pyruvate dehydrogenase deficiency, there is a backup of pyruvate and alanine resulting in lactic acidosis. This can be caused from a congenital enzyme deficiency or can be acquired due to vitamin B1 deficiency (in alcoholics for example). The treatment for this is to increase the intake of ketogenic nutrients. The two ketogenic amino acids are leucine and lysine (choice D). The other amino acids listed here are not ketogenic.

Histamine (choice C) is not an amino acid and is unlikely to be an option on well-edited exam.
A prisoner goes on a hunger strike to protest the conditions of his detainment. After several days, his blood glucose level is significantly reduced. In order to keep a constant supply of energy to his brain and muscle, which of the following molecules is his liver releasing?

a. Beta-hydroxybutyrate  
b. Fatty acids  
c. Glucose-6-P  
d. Glycogen  
e. Nucleic acids

**Correct answer:** A

The liver's role during starvation is to release both glucose (via gluconeogenesis) and ketone bodies into the blood. Adipose tissue provides fatty acids both for muscle and liver. Muscle oxidizes the fatty acids completely to CO2 and water for energy. Liver converts about half of the fatty acids it takes up into ketone bodies (acetoacetate and b-hydroxybutyrate), which it releases into the blood. The other half is oxidized to provide energy for the liver.

Glycogen reserves are gone after 30 hours, but even when present, glycogen is never released into the blood as a polymer. It is broken down to G1P, which is then converted to G6P, which is then hydrolyzed to free glucose. Only free glucose can exit the cell.

Nucleic acids are never used as fuels, making "E" a ridiculous distractor. Better options would have been: amino acids, glycerol, lactate.
A 2 year old female presents with a new onset pain in her right thigh. When reviewing her medical history you find that she has had one episode of aspergillus pneumonia, recurrent impetigo secondary to staphylococcus aureus infection, and severe diaper rash secondary to candidiasis infection. You suspect osteomyelitis but are also concerned about a more serious underlying etiology that has caused these recurrent infections. Which test would most likely lead to a diagnosis?

a. Flow cytometry using CD11 and CD18 monoclonal antibodies
b. Myeloperoxidase staining of neutrophils
c. Nitroblue tetrazolium test
d. Sweat chloride test
e. T cell and absolute lymphocyte count

**Correct answer:** C

This child most likely has chronic granulomatous disease, which is a deficiency in NADPH oxidase which is the initial step in the respiratory burst present in the phagolysosome. CGD usually presents with recurrent bacterial and fungal infections before the age of five. The test of choice for CGD is nitroblue tetrazolium test (choice C). This tests for the respiratory burst found in neutrophils which would be absent in CGD.

Flow cytometry using CD11 and CD18 monoclonal antibodies (choice A) would be used in diagnosing leukocyte adhesion deficiencies. A typical presentation for these disorders would be a delay in the separation of the umbilical cord at birth.

Myeloperoxidase staining of neutrophils (choice B) is used to diagnose myeloperoxidase deficiency. Although myeloperoxidase exists in the same respiratory burst as NADPH oxidase, patients with myeloperoxidase deficiency are usually clinically normal.

Sweat chloride test (choice D) is used to diagnose cystic fibrosis. Although patients with CF are more likely to develop pneumonia, the pathogens would more likely be bacterial rather than fungal.

T cell and absolute lymphocyte count (choice E) would be useful in the diagnosis of severe combined immunodeficiency syndrome. This would present early in life with failure to thrive and severe life-threatening infections.
A 3-year-old boy has suffered from recurrent bouts of pneumonia and soft tissue abscesses since birth. He currently experiences fever, chills, dyspnea, and irritability. A sputum culture is taken, which grows out Aspergillus. His pediatrician decides to do a nitroblue-tetrazolium (NBT) test, which is negative. Which enzyme deficiency is likely responsible for this boy’s condition?

- a. Glucose-6-phosphate dehydrogenase (G6PD)
- b. Glutathione reductase
- c. Myeloperoxidase
- d. NADPH oxidase
- e. Superoxide dismutase

correct answer: D

This boy suffers from chronic granulomatous disease (CGD), which is a congenital immunodeficiency caused by a deficiency in NADPH oxidase (choice D). The enzyme is responsible for reducing molecular oxygen to produce superoxide free radical, which is the first step in the oxygen-dependent respiratory burst used by phagocytes to kill bacteria. Patients with this condition get recurrent infections, including pneumonia, abscesses, and suppurative arthritis. The NBT test is negative in patients with CGD, since their phagocytes can’t produce superoxide to reduce NBT.

Glucose-6-phosphate dehydrogenase (G6PD, choice A) is an enzyme that generates NADPH, used in fatty acid synthesis in many tissues, and by glutathione reductase to prevent oxidative damage in red blood cells. G6PD deficiency commonly manifests as acute hemolytic anemia in response to infections, certain medications, or fava beans.

Glutathione reductase (choice B) "regenerates" the cellular antioxidant glutathione by transforming oxidized GSSC to the reduced form GSH. If glutathione reductase is deficient, the oxidative stress can lead to cell lysis. However, a congenital deficiency of this enzyme is exceedingly rare.

Myeloperoxidase (choice C) is a lysosomal enzyme found in neutrophils, and is also involved in the oxygen-dependent respiratory burst. Screening for a myeloperoxidase deficiency involves immunochemical staining for peroxidase.

Superoxide dismutase (SOD, choice E) catalyzes the dissociation of the reactive oxygen species superoxide into oxygen and hydrogen peroxide. Mutations in SOD1 have been linked to familial amyotrophic lateral sclerosis (ALS).
A 56 year old alcoholic presents to the ER with multiple neurological problems and history of multiple ER visits as seen when you get his chart from the nurse. The nurse sets him up with multiple fluids of which one is thiamine. If Thiamine deficiency is presumed to be the reason for his presentation which of the following enzymes is most likely to be abnormally functioning?

a. Dopamine hydroxylase
b. Propionyl CoA carboxylase
c. Pyruvate carboxylase
d. Pyruvate dehydrogenase
e. Thymidylate synthase

correct answer: D
Thiamine is involved with dehydrogenase reactions (PDH and alpha ketoglutarate dehydrogenase for TCA cycle); and transketolase for the HMP shunt. Vitamin C serves as a coenzyme for dopamine hydroxylase responsible for catecholamine synthesis. Folic Acid is involved with thymidylate synthase. Biotin is involved with the carboxylase reactions (including pyruvate carboxylase in gluconeogenesis, acetyl CoA carboxylase in fatty acid synthesis, and propionyl CoA carboxylase for Odd-carbon FA, Val, Met, Ile, pathways).

In summary,
Biotin for carboxylase reactions.
Thiamine for dehydrogenase reactions and transketolase.
B9(folate) for transferases.
Vit C for hydroxylases.
B12 classically for homocysteine methyltransferase and methmalonyl CoA mutase.
Kartagener’s Syndrome results from a defect in the arm of the dynein molecule, and may produce such symptoms as bronchiectasis and recurrent sinus infections. Which of the following is also a common consequence of Kartagener’s Syndrome?

a. Elastic Skin  
b. Infertility  
c. Interstitial Fibrosis  
d. Tendon Xanthomas  
e. Tracheoesophageal Fistula

**Correct answer:** B

A defect in the dynein protein causes dysfunction of ciliary movement, and manifests in patients with symptoms related to the improper beating of cilia. Bronchiectasis and recurrent sinus infections result from the inability to adequately remove particulate matter and infectious materials from the nasopharynx and bronchial trees. Importantly, dysfunction in cilia motility results in both male and female infertility (due to deficits in ovary migration and sperm flagella).

Elastic skin (A) (Ehler’s Danlos Syndrome) results from faulty collagen synthesis and the accumulation of tropocollagen in the extracellular space.

Interstitial fibrosis (IF) (C) involves bronchial inflammation and scarring and is unrelated to overt ciliary dysfunction. IF often results from asbestosis, silicosis and other pneumoconioses.

Tendon Xanthomas (D) are not related to ciliary function at all, but are a common symptom of familial hypercholesterolemia.

TE fistulas (E) occur when the lower esophagus joins the trachea, and leaves the upper esophagus as a blind pouch. They are a product of improper development of the tracheoesophageal junction, often associated with polyhydramnios during pregnancy.
A tall 18 year old patient is brought by his mother to his pediatrician office complaining of chest pain. On questioning, his mother tells the doctor that her son goes to a school for the mentally retarded due to a low IQ. On examination the patient slouches, and has blurry vision, and EKG reveals ST changes on V1-4. A deficiency of which product is the cause of the symptoms involved with this disorder?

a. Fibrillin gene
b. Type III collagen.
c. Cystathionine synthase
d. Pyruvate kinase
e. Glucose-6-Phosphatase

**Correct answer:** C

Homocystinuria, also known as Cystathionine beta synthase deficiency, is an inherited disorder of the metabolism of the amino acid methionine, often involving cystathionine beta synthase. It is an inherited autosomal recessive.

Symptoms include: Tall stature, mental retardation, kyphosis, ectopia lens, antherosclerosis.

Answer (A) Fibrillin is a gene defect in Marfan's syndrome. Although clinically, this case resembles Marfan's syndrome, this patient is mentally retarded, which excludes the diagnosis.

Answer (B) Type III collagen deficiency is related to Ehlers-Danlos syndrome.

Answer (D) A deficiency in pyruvate kinase will cause hemolytic anemia since the RBC has no mitochondria and with a deficiency of pyruvate kinase the RBCs have no way of making ATP.

Answer (E) A deficiency of glucose-6-phosphatase is related to von Gierke's disease which is a glycogen storage disease characterized by loss of blood homeostasis and glycogen and lipid metabolism.
A tall 18 year old patient is brought by his mother to your office complaining of chest pain. On examination he is mildly retarded, slouches, and has blurry vision. His EKG reveals ST changes on V1-4. What is the deficient product involved with this disorder?

a. Fibrillin gene  
b. Type III collagen.  
c. Cystathionine synthase  
d. Pyruvate kinase  
e. Glucose-6-Phosphatase

**Correct answer: C**

Homocystinuria, also known as Cystathionine beta synthase deficiency, is an inherited disorder of the metabolism of the amino acid methionine, often involving cystathionine beta synthase. It is an inherited autosomal recessive.

Symptoms include: Tall stature, mental retardation, kyphosis, ectopia lens, antherosclerosis.

Answer (A) Fibrillin is a gene defect in Marfan's syndrome. Although clinically, this case resembles Marfan's syndrome, this patient is mentally retarded, which excludes the diagnosis.

Answer (B) Type III collagen deficiency is related to Ehlers-Danlos syndrome. Answers (D) and (E) are incorrect.
A 7 year old girl was seen in a local hospital with complaints of crampy abdominal pain. Her two younger siblings had not been sick, and her parents were not aware of any sick contacts at school. Physical examination showed diffuse, mild abdominal tenderness. Imaging studies later revealed small bowel bowel obstruction. Thorough workup of this patient in the ER included a urinalysis, which showed the following results:

- Leukocyte esterase: negative
- Nitrites: negative
- Glucose: negative
- Ketones: negative
- Protein: negative
- Specific gravity: 1.055

Further laboratory studies revealed elevated blood and urine concentrations of fructose. The enzyme deficient in this patient directly facilitates the production of which of the following products:

a. Fructose-1-P
b. Fructose-6-P
c. Dihydroxyacetone-P
d. Glyceraldehyde-3-P
e. Glyceraldehyde

**Correct answer:** A

The patient in question has an incidental finding of essential fructosuria, which is caused by deficiency of fructokinase. Fructokinase converts fructose to Fructose-1-P, so fructokinase deficiency results in the accumulation of fructose in the serum and urine.

Fructose-6-P is the substrate of the rate-limiting enzyme phosphofructokinase in the glycolytic pathway and is not involved in essential fructosuria, so (B) is incorrect.

Do not confuse essential fructosuria with the more severe condition of fructose intolerance, which involves the intracellular accumulation of Fructose-1-P due to Aldolase B deficiency. Aldolase B cleaves Fructose-6-P into Dihydroxyacetone-P (C) and Glyceraldehyde (E), both of which can be further metabolized to Glyceraldehyde-3-P (D).
A 55-year-old man who has smoked for the past 35 years is experiencing seizures. Before today he has never had any seizures. His plasma osmolarity measures 250 mOsm/kg. An MRI is taken and shown above. MRI shows generalized cerebral edema but no lesions. Which of the following minerals is most likely responsible for the signs and symptoms in this patient?

a. Calcium  
b. Magnesium  
c. Phosphorus  
d. Potassium  
e. Sodium

correct answer: E

Sodium is the primary mineral that contributes to the plasma osmolarity. A low plasma osmolarity almost always indicates the presence of hyponatremia because blood glucose and BUN are less important contributors. Alterations in serum sodium concentration establish an osmotic gradient between ECF and ICF. This gradient results in movement of water into the ICF in order to equalize the osmolarity in both compartments, which accounts for the cerebral edema that is causing the seizures. Ectopic secretion of ADH by small cell carcinoma accounts for the hyponatremia.

Calcium (Choice A) is incorrect; calcium is important in mineralization of bone and teeth, nerve conduction, muscle contraction, signal transduction across membranes among other functions.

Magnesium (Choice B) is incorrect; magnesium is involved in calcium metabolism, muscle contraction, and nerve impulse propagation.

Phosphorous (Choice C) is incorrect; phosphorous is an important component of DNA, RNA, ATP, and phosphorylated vitamins.

Potassium (Choice D) is incorrect; potassium only plays a minor role in the control of oncotic pressure in the ECF compartment.
A 35 year old African-American woman presents complaining of vision problems, fatigue, and constipation. She reports that “her doctor has told her that she has kidney stones.” Her skin examination revealed tender red nodules. Overexpression of which enzyme is consistent with her presentation?

- a. Desmolase
- b. Alkaline phosphatase
- c. Adenylate cyclase
- d. Sodium-potassium ATPase
- e. 1-alpha hydroxylase

**Correct answer:** E

This patient most likely has sarcoidosis. African Americans are at higher risk for this disease compared to the general population. Her symptoms are consistent with hypercalcemia, which may cause CNS symptoms. The skin rash described in the case is erythema nodosum, and the vision problems are most likely related to granulomas in the optic nerve. These findings should raise a suspicion of sarcoidosis. The granulomas have elevated levels of 1-alpha hydroxylase, an enzyme normally found in the renal proximal tubules that is responsible for the final step in biosynthesis of Vitamin D3. Elevated levels of Vitamin D cause hypercalcemia by increasing calcium’s absorption in the gut and decreasing its excretion by the kidneys.

Desmolase is the first step in the conversion of cholesterol to pregnenolone.

Alkaline phosphatase is important in the biliary tree indicating biliary blockage. It is also found in bones.

Adenylate cyclase is ubiquitous inside cells and can lead to phosphorylation of various enzymes. Although it can lead to increased intracellular calcium, it does not present the classic picture of sarcoidosis.

Na/K ATPase is responsible for the active transport of sodium and potassium across the cell membrane to maintain the resting potential and adequate cell volume.
A 6 year old male child is brought into the pediatrics clinic. According to his parents he has been having diarrhea and a red rash. They also report that at times, he is “somewhat uncoordinated in his walking.” A thorough evaluation does not reveal any nutritional deficiencies. Which of the following do you expect to see?

- a. Elevated neutral amino acid in his urine
- b. Reduced levels of tryptophan in his urine
- c. Lead poisoning
- d. A history of viral infection in the gastrointestinal tract
- e. An autosomal dominant condition upon genetic analysis

**correct answer:** A

This patient has Hartnup disease, an autosomal recessive condition (E is incorrect) characterized by a poor absorption of neutral amino acids (such as tryptophan) due to a defective sodium-dependent and chloride-independent transporter mainly found in the kidney and the gut. Given that the patient ingests those amino acids, as suggested by normal nutrition, he will have aminoaciduria, as he cannot absorb tryptophan (B is incorrect). Therefore these patients present with a pellagra like syndrome, characterized by diarrhea, rash, and neurologic symptoms, such as intermittent ataxia.

C is incorrect. Lead poisoning may cause neurologic damage, microcytic anemia, basophilic stippling of red blood cells, as well as a bluish line along the gums (Burton’s line). The case does not report exposure to lead. A skin rash is not commonly associated with lead poisoning.

D is incorrect. Although viral gastroenteritis could explain the diarrhea, it does not explain the totality of the patient problems.
A Southeast Asian immigrant child is noted to be severely retarded. Physical examination reveals a pot-bellied, pale child with a puffy face. The child’s tongue is enlarged. Dietary deficiency of which of the following substances can produce this pattern?

a. calcium  
b. Iodine  
c. iron  
d. Magnesium  
e. Selenium

**Correct answer:** B

The correct answer is B. The disease is cretinism, characterized by a profound lack of thyroid hormone in a developing child, leading to mental retardation and the physical findings described in the question stem. Cretinism can be due to dietary deficiency of iodine (now rare in this country because of iodized salt), to developmental failure of thyroid formation, or to a defect in thyroxine synthesis.

Calcium deficiency (choice A) in children can cause osteoporosis or osteopenia.

Iron deficiency (choice C) can cause a hypochromic, microcytic anemia.

Magnesium deficiency (choice D) is uncommon, but can cause decreased reflexes, and blunts the parathyroid response to hypocalcemia.

Selenium deficiency (choice E) is rare, but may cause a reversible form of cardiomyopathy.
A 64 year old male presents with his wife to his primary care physician complaining of diarrhea, a rash on his arms and legs, and severe flushing of the skin of his upper thorax and head. The patient's wife relays a history of congestive heart disease and small intestine cancer 6 years ago for which the patient received radiation therapy. Urinalysis reveals significantly decreased levels of N'\-methylnicotinamide. A diagnosis of pellagra is made. Which of the following is most likely the primary cause of niacin deficiency in this patient?

a. Adverse effect of digoxin therapy  
b. Adverse effect of statin therapy  
c. Decreased absorption due to a complication of radiation therapy  
d. Carcinoid Syndrome  
e. Celiac disease

correct answer: D

Carcinoid syndrome is a complication of carcinoid cancer and is characterized by abnormal secretion of serotonin (5-ht). Flushing and gasterointestinal disturbances are the the most common symptoms associated with carcinoid syndrome. This patient has a history of small intestinal cancer, a common cause of carcinoid syndrome. The increase in serotonin production causes a deficiency of tryptophan, which is a neccesary precursor for niacin synthesis.
One of the proposed mechanisms for the development of problems in the eyes and kidneys of diabetics involves the formation of excess sorbitol from glucose. Excess sorbitol in these organs can be toxic and is known to form cataracts upon buildup within the lens of the eye. The enzyme that performs this reaction is:

a. Aldose Reductase  
b. Galactose  
c. Glucokinase  
d. Hexokinase  
e. Sorbitol Dehydrogenase

**Correct answer:** A

Aldose reductase performs the following reaction:

\[
\text{Glucose} \rightarrow \text{Sorbitol}
\]

This reaction is part of the polyol pathway utilized to synthesize fructose within the human body. The levels of Aldose Reductase are typically quite low, however in response to chronic hyperglycemia (as seen in diabetics) the levels can increase, thus increasing the levels of sorbitol.
Severe Combined Immunodeficiency Disease (SCID) can result from a deficiency of this enzyme involved in purine catabolism, which results in inhibition of ribonucleotide reductase.

a. Adenosine Deaminase  
b. Dihydrofolate Reductase  
c. HGPRT  
d. Thymidylate Synthase  
e. Xanthine Oxidase

**Correct answer:** A  
This is one of the most common causes of SCID (a deficiency in both B and T lymphocytes). ADA deficiency disrupts the balance of the nucleotide pools and causes indirect inhibition of ribonucleotide reductase. Ribonucleotide Reductase is essential for DNA replication as it forms deoxyribonucleotides.
The membrane protein Bax forms pores in the membranes of mitochondria. These pores are normally blocked by the protein Bcl2. When a cell undergoes apoptosis, proteins bind to Bcl2, removing it from Bax. This allows for the release of a molecule that triggers apoptosis. The molecule implicated here is:

- a. AMP
- b. ATP
- c. Cytochrome C
- d. Mitochondrial DNA
- e. Pyruvate

**Correct answer:** C

Leakage of cytochrome C into the cytoplasm of a cell triggers Apaf-1 to form apoptosome which signals a cascade of caspases and proteins are degraded. In some cancers excess Bcl2 is formed, preventing apoptosis from occurring.
A newborn female has a large and distorted cranium, short and deformed limbs, and very blue scleras (whites of the eyes). Radiographs demonstrate multiple limb fractures and suggest a diagnosis of osteogenesis imperfecta (brittle bone disease-155210). Analysis of type I collagen protein, a triple helix formed from two 1 and one 2 collagen chains, shows a 50% reduction in the amount of type I collagen in the baby's skin. DNA analysis demonstrates the presence of two normal 1 alleles and one normal 2 allele. These results are best explained by which of the following?

a. Deficiency of alpha 1 collagen peptide synthesis
b. Inability of alpha 1 chains to incorporate into triple helix
c. Defective alpha1 chains that interrupt triple helix formation
d. Incorporation of defective alpha 2 chains that cause instability and degradation of the triple helix
e. A missense mutation that alters the synthesis of alpha 1 chains

Correct answer: D

Collagen peptides assemble into helical tertiary structures that form quaternary triple helices. The triple helices in turn assemble end to end to form collagen fibrils that are essential for connective tissue strength. Over 15 types of collagen contribute to the connective tissue of various organs, including the contribution of type I collagen to eyes, bones, and skin. The fact that only one of two 2 alleles is normal in this case implies that a mutant 2 allele could be responsible for the disease (even if the 2 locus is on the X chromosome, since the baby is female with two X chromosomes). The mutant 2 collagen peptide would be incorporated into half of the type I collagen triple helices, causing a 50% reduction in normal type I collagen. (A mutant 1 collagen peptide would distort 75% of the molecules because two 1 peptides go into each triple helix.) The ability of one abnormal collagen peptide allele to alter triple helix structure with subsequent degradation is well-documented and colorfully named protein suicide or, more properly, a dominant-negative mutation.
A teenage boy presents to clinic complaining of muscle cramps on exercise. Past history indicates he had some coordination problems in childhood and received occupational therapy. Blood tests show an increased amount of lactic acid at rest, with dramatic increases on exercise testing. Simultaneous measures of capillary oxygenation by a surface probe were normal. The abnormality most likely involves which of the following?

- a. Glycolysis in the lysosomes
- b. Glycolysis in the cytosol
- c. Respiratory chain in the mitochondria
- d. Glycogen breakdown in the mitochondria
- e. Gluconeogenesis in the lysosomes

**Correct answer:** C

Under conditions of plentiful oxygen (aerobic metabolism), pyruvate formed from glycolysis in the cytosol is metabolized to acetylCoA. Acetyl CoA enters the mitochondria and the citric acid cycle in the conversion of oxaloacetate to citrate, generating NADH and FADH2 reducing equivalents that generate ATP through oxidation in the mitochondrial respiratory chain. In respiratory chain disorders, the disrupted electron transport chain does not function as well, causing more pyruvate to be converted to lactate in muscle with muscle weakness and cramping. Lactate from exercising muscle is normally converted to glucose by the liver, but excess lactate produced in severe respiratory chain disorders accumulates in serum to lower the pH (acidosis). Mitochondria are called the powerhouses of the cell since they contain the citric acid cycle and the respiratory chain that generates abundant ATP through oxidative phosphorylation.
A neonate tests positive for elevated serum phenylalanine during a routine screening test. The child is started on a phenylalanine restriction diet. Despite complete compliance with the diet, the mother still notices a “mousy” smell to the child’s sweat, and the child still develops severe neurological defects. Which of the following neurotransmitters is likely to have been synthesized at a reduced level in the child’s CNS?

a. Acetylcholine
b. Gamma-aminobutyric acid
c. Glutamine
d. Norepinephrine
e. Histamine

correct answer: D

There are two approaches to this question. Using the buzzword approach, one must recognize this as a case of phenylketonuria (PKU) by the increased serum phenylalanine and the “mousy” smell to the child’s sweat. This is necessary, but not sufficient for answering this question. If one continues to use the “buzzword memorization” approach for the rest of this question, though, you would have to remember that there are various genetic mutations that can lead to hyperphenylalaninemas, including mutations that directly affect the enzyme phenylalanine hydroxylase and mutations in the pathway that alter the synthesis or recycling of that enzyme’s cofactor, tetrahydrobiopterin (BH4), and then all the pathways associated with those enzymes. This approach is not recommended.

What is recommended is a general principles approach. This approach also requires memorization, but it requires memorization of general concepts. In biochemistry questions like this one, you do need to memorize the molecules in key biochemical pathway that appear in a variety of places in medicine. Here you should know:

Phenylalanine --> Tyrosine --> L-Dopa --> Dopamine --> Norepinephrine --> Epinephrine

Or at least that all 3 of the catecholamines (dopamine, norepinephrine, and epinephrine) are derived from the amino acid tyrosine. As was stated before, you do need to recognize this as a case of PKU (it is likely a case related to a mutation of BH4 synthesis or recycling since dietary control doesn’t help, but you don’t need to know that). The only thing you really do need to know about PKU is how to recognize it, and that the primary problem, no matter what mutation is leading to the phenotype, is the inability to synthesis tyrosine from phenylalanine. This leads to an excess of neurotoxic phenylalanine and its secondary metabolites (like the keto forms that give PKU its name) building up in the body fluids (blood, urine, sweat, and CNS). This idea points out to another general concept about all “inborn errors of metabolism”, of which PKU is one. Often times it is not the inability to synthesize something that is the problem (although in this case it was), but rather the inability to get rid of something that has the potential to be toxic that is the problem in “inborn errors of metabolism” (or toxicities of drugs that block biosynthesis pathways for that matter). All the other answers are wrong simply because their synthesis does not require tyrosine as a precursor or the use of tetrahydrobiopterin as a cofactor (note: serotonin was not an answer choice, because its synthesis also requires BH4, and could also have been a correct answer, but you need not memorize that either).

Thompson and Thompson Genetics in Medicine, 6th ed., pp. 206-210
A 68 year old male presents with tremor at rest, bradykinesia, stooped posture and shuffling gait. The amino acid that is the precursor for the substance deficient in this patient is:

- a. Glycine
- b. Glutamate
- c. Glucosamine
- d. Tyrosine
- e. Tryptophan

**Correct answer:** D

Described above are typical symptoms of parkinsonism (not to be confused with Parkinson’s Disease, which is the main cause of parkinsonism). Parkinsonism is related to a deficiency in the neurotransmitter dopamine. Tyrosine 3-monoxygenase (tyrosine hydroxylase) hydroxylates L-Tyrosine converting it to L-Dopa. Aromatic L-amino acid decarboxylase (also known as dopa-decarboxylase) decarboxylates L-Dopa to Dopamine. Dopamine can be further processed to norepinephrine and epinephrine.

Tryptophan is converted to 5-hydroxytryptophan (5-HT) by tryptophan hydroxylase. 5-HT is decarboxylated by aromatic L-amino acid decarboxylase to serotonin (5-hydroxytryptamine or 5-HT).

Glycine is a precursor of porphyrin and is itself an inhibitory neurotransmitter in the CNS. Parkinsonism, however, is unrelated to glycine.

Glutamate is also a neurotransmitter (the most abundant excitatory neurotransmitter) and a precursor for the inhibitory neurotransmitter GABA through glutamate decarboxylase.

Glucosamine is not an amino acid.
A 27 year old woman presents with complaints of abdominal pain. On physical exam, she screams in pain when you palpate the right lower quadrant of her abdomen. Laboratory workup reveals a positive beta-hCG. She acknowledges a complete STD test done 5 days ago, the results of which was "negative for everything". She has not engaged in sexual activity since receiving her test results. When asked about her past medical history, she endorses chronic recurring respiratory and ear infections as a child. Abnormalities of which of the following structures explain this patient’s problems?

a. Plasma membrane proteins
b. Microtubules doublets surrounding a central pair
c. Phagolysosome formation
d. Inflammation of a specific structure in the intestinal tract
e. None of the above

**Correct answer: B**

This patient most likely has Kartagener syndrome (KS), also called primary ciliary dyskinesia, in which cilia are non-motile. Cilia are hair-like projections attached to the cell body, covered by plasma membrane. KS is a rare autosomal recessive genetic disorder characterized by a defect in the action of the cilia lining the respiratory tract (lower and upper, sinuses, Eustachian tube, middle ear) and fallopian tube. In the respiratory tract, cilia move debris through a beating upward motion, and those debris are coughed up. Accumulation causes infection. In the fallopian tube, cilia function to move the egg to the uterus for implantation. Cilia are comprised of cytoskeletal proteins: 9 pair of microtubules made of dynein molecules surrounded by a central bridge of dynein. Dysfunctional cilia cause the egg to grow in the fallopian tube, causing an ectopic pregnancy. A very common symptom of ectopic pregnancy is pain in the lower quadrant. Ectopic pregnancy can also be caused by pelvic inflammatory disease, particularly in patients with a history of chlamydial infection. The fact that she had a negative STD panel negates that possibility. Additionally, she has a positive beta-hCG, confirming a pregnancy.

Answer A is incorrect; cilia are hair-like projections, of the cell. They are covered by plasma membrane; the actual pathology is explained by cytoskeletal abnormality, not plasma membrane proteins.

Answer C is incorrect; the formation of phagolysosomes does not require cilia. The phagosome and the lysosome are 2 cellular organelles, which function independently of cilia.

Answer D is incorrect; although appendicitis causes lower quadrant pain, the clinical picture is more consistent with Kartagener syndrome.
An experiment is conducted in which researchers are investigating the effect of various substances during the menstrual cycle. Substance X was found to directly stimulate expression of growth factors involved in blood vessel formation. Which of the following is most likely to be substance X?

- a. Luteinizing hormone
- b. Androstenedione
- c. Follicle stimulating hormone
- d. Estrogen
- e. Progesterone

**correct answer: E**

Progesterone is a steroid hormone secreted by the gonads, the placenta, as well as the brain. It prepares the reproductive tract for a pregnancy (think progesterone = pro-gestation), stimulates development of the spiral arteries in the endometrium by promoting the expression of angiogenic factors (angiopoietin-1, vascular endothelial growth factor (VEGF), fibronectin). As the main hormone of the secretory phase of ovulation, it causes endometrial glands to enlarge and secrete glycogen, a essential nutrient for the thickened uterus. If no fertilization occurs, the spiral arteries undergo spasms and necrosis, and then disintegrate in the stratum functionalis.

Answer A is incorrect; although luteinizing hormone stimulates the release of progesterone, it does not directly stimulate the endometrial events (vessel formation, gland enlargement)

Answer B is incorrect; androstenedione is an intermediate in the pathway of testosterone synthesis

Answer C is incorrect; follicle stimulating hormone is released by the anterior pituitary, and stimulates the formation of estrogen by increasing the expression of aromatase in the gonads and other tissues in the body (brain, skin, bone, adipose, etc)

Answer D is incorrect; estrogen is the main hormone in the proliferate phase. The influence of estrogen peaks right before ovulation. After ovulation, levels of estrogen reduce and the corpus lustrum starts to make progesterone, which marks the secretory phase.
A 67 year-old male presents with hemoptysis and cough for the past 3 months. He also has night sweats but no weight loss. He is afebrile with an unremarkable physical exam. His chest X-ray shows three cavitary lesions all in the upper left lung lobe. His treatment regimen consists of isoniazid, rifampin, pyrazinamide and ethambutol. After several months of treatment his pulmonary symptoms have decreased but the patient states he has new tingling sensations in his hands and feet. He does not have diabetes mellitus.

At this point you realize that you forgot to supplement his drug regimen with which of the following?

a. Iron (ferrous sulfate) because rifampin can cause pernicious anemia
b. Vitamin B6 because isoniazid decreases its absorption
c. Vitamin B12 because isoniazid decreases its production
d. Vitamin D because isoniazid may decrease its production in the kidney
e. Folate because rifampin may inhibit its absorption

**correct answer: B**

The correct answer is B. The patient has active tuberculosis (pulmonary) and is begun on a trial of RIPE antibiotics. Later, the patient presents with peripheral neuropathy. Isoniazid (INH) decreases the absorption of vitamin B6 and thus this vitamin must be given concurrently with isoniazid. INH may also cause hepatitis, and so liver function should be tested before drug administration and during treatment.

Choice A is incorrect; iron deficiency would not cause pernicious anemia. Pernicious anemia is caused by an autoimmune process that destroys parietal cells of the stomach and intrinsic factor, reducing the absorption of vitamin B12 and leading to macrocytic anemia.

Choice C is incorrect; B12 is not produced in the human body rather it is absorbed

Choice D is incorrect; isoniazid does not affect vitamin D production in the kidney.

Choice E is incorrect; rifampin does not inhibit folate absorption.
Vitamin C deficiency results in scurvy. Vitamin C functions as a co-factor in which of the following steps of the collagen synthesis cascade?

- a. Cleavage of procollagen
- b. Exocytosis of collagen
- c. Glycosylation of procollagen
- d. Hydroxylation of preprocollagen
- e. Self-assembly of procollagen

**Correct answer:** D

The hydroxylation of specific proline and lysine residues on the preprocollagen strand requires vitamin C as a cofactor in the endoplasmic reticulum. There, the pro-alpha chain of collagen is glycosylated to form the triple-helixed procollagen. Procollagen is then sent to the Golgi apparatus and is exocytosed from the cell, where the ends are cleaved to form tropocollagen. These molecules are cross-linked to form collagen fibrils. Unlike most proteins, post-translational modification occurs in the RER rather than the Golgi apparatus.
A 4-month-old infant is brought by his parents to the ER where imaging shows a fractured rib and a subdural hematoma; your resident subsequently suspects child abuse. However, on further physical examination you note that the infant appears to be mentally retarded and his hair is lacking color and appears abnormally twisted. Labs are significant for an abnormally low copper level of 4.3 nM. It is found that the infant has Menkes disease, an abnormal functioning of lysyl oxidase due to decreased copper that normally serves as a cofactor in the cross-linking of collagen fibrils. Which of the following amino acids is most likely related to the mechanism of this infant’s hypopigmented hair?

a. Arginine  
b. Glutamate  
c. Histidine  
d. Tryptophan  
e. Tyrosine

**Correct answer:** E

Phenylalanine gives rise to tyrosine which gives rise to DOPA, which is a precursor of the pigment melanin (responsible for pigmentation of skin and hair). DOPA can also give rise to dopamine (via decarboxylation) and thereafter the catecholamines (norepinephrine, then epinephrine). Therefore, this is a critical biochemical pathway. In Menkes disease (X-linked recessive inheritance), decreased copper levels lead to a number of problems. Copper is a cofactor for dopamine beta hydroxylase (hence the disruption in the pathway that leads to melanin synthesis), in addition to lysyl oxidase. Brain MRI’s would show impaired myelination (due to decreased cytochrome C oxidase activity); diffuse atrophy from neuronal degeneration and gliosis; ventriculomegaly; and tortuous vessels (due to decreased lysyl oxidase activity).

(A) Arginine gives rise to creatine, urea, nitric oxide (NO).  
(B) Glutamate gives rise to GABA and glutathione.  
(C) Histidine gives rise to histamine.  
(D) Tryptophan gives rise to niacin → NAD+/NADP+; and serotonin → melatonin.
A 2 months old infant was brought to your office with an obvious depigmentation and multiple skin lesions. Upon further examination the infant appear to have tortuous arteries. The doctor suspect Menke's disease. What is mechanism behind this disease?

a. The mother did not consume enough copper
b. The infant diet did not contain any copper
c. The intestinal epithelial cells did not absorb copper
d. The intestinal epithelial cells cannot efflux absorbed copper
e. Idiopathic

**Correct answer:** D

This scenario is typical for menke's disease. Characterized by skin depigmentation and tortuous arteries. The biochemistry behind this question is the lack of cross linking due to copper deficiency. The copper is absorbed normally from by the epithelial cells but there is a inability to efflux the copper to different organs leading to lack of melanin and depigmentation.
What percentage of Down’s syndrome patients also have congenital cardiovascular disease?

a. 1%
b. 5%
c. 20%
d. 50%
e. 90%

**Correct answer:** C

The correct answer is C. This fact is worth remembering: one-fifth of Down's syndrome patients have congenital cardiovascular disease, most commonly an ostium primum type of ASD and/or a ventricular septal defect. This is a sufficiently high incidence to justify at least one echocardiogram in each of these children's lives. Affected children are also particularly vulnerable to seizures, and as adults may develop an Alzheimer-like dementia in their mid 40s.
A study is conducted to ascertain the risk factors contributing to asthma in Bronx youth ages 9-12. 422 Bronx residents, ages 9-12, have agreed to participate in the study. Given the study’s objective, what is the best approach to use?

- a. Case-control
- b. Cohort
- c. Clinical trial
- d. Meta-analysis
- e. Randomized, double-blind trial

**Correct answer:** A

Since the participants are already in the target range, the best choice to determine potential risk factors is a retrospective case-control study (perform histories and determine the odds ratios for each risk factor). A cohort study would involve younger children being grouped by a risk factor one wishes to test and then determining the incidence of asthma over time in each of the groups. Clinical trials require some sort of intervention/therapy, and meta-analysis refers to aggregating other studies/analyses to bolster a given conclusion.
A 24-year-old male visits a clinic for an HIV test. When he returns three days later for the result, the nurse informs him that the test was positive and a second test is being performed to validate the result. The young man decides to investigate what the likelihood is of the first test being incorrect. What statistical value below would answer this young man’s question?

a. Negative predictive value (NPV)
b. Positive predictive value (PPV)
c. Prevalence
d. Relative risk
e. Sensitivity

Correct answer: B

The value he is searching for is the false positive rate. When confronted with test vs. disease questions, it is best to draw a 2x2 table: Row 1: a (++), b (+-); Row 2: c (-+), and d (--). For example, b is (+-), meaning the test is positive but person does not have the disease. To determine b—the false positive rate—one can take either (1-PPV) = 1 - a/(a+b) = b/(a+b) [% of false positives] or (1-Specificity) = 1 - d/(d+b) = b/(d+b) [% of false positives]. Therefore, the answer is both PPV and specificity. Sensitivity = a/(a+c). NPV = d/(c+d).
A fictitious disease, RomeOnHBOFever, is slowly taking over the entire NY medical student community (3,000 students). In 2006, the prevalence of this disease within this community was 10%. In 2007, 270 new cases of RomeOnHBO Fever were reported. What is incidence of this gripping disease in 2007?

- a. 5%
- b. 9%
- c. 10%
- d. 15%
- e. 19%

**Correct answer:** C

Incidence = (# of new cases) / (# at risk). For 2007, there are 270 new cases and 2700 people at risk; therefore, the incidence is 10%. Do not forget to limit the incidence value to only those at risk. The 300 infected from 2006 (10% of 3000) are no longer at risk for the disease--they already have it!
Investigators are studying the use of a new laboratory test to identify patients with a particular disease. The table above summarizes the results of initial research involving 200 subjects. Which of the following is the approximate sensitivity of a positive test result?

- a. 0.30
- b. 0.33
- c. 0.60
- d. 0.67
- e. 0.75

**correct answer:** E

Sensitivity, as defined in biostatistics, is the number of true positive results divided by the total positive test results (true positive plus false negative).

Sensitivity = 60/(60+20) = 0.75
A field researcher is conducting a study of HIV transmission in South Africa. She has already obtained access to a cohort of HIV-positive patients and a cohort of age-matched HIV-negative patients. She now plans to undertake a case-control study using the data set. Which of the following would be a feasible outcome measure for this study?

a. The average death rate between cohorts
b. The frequency difference of past unprotected sex between cohorts
c. The presence of HIV in children of HIV-positive enrollees in the next 5 years
d. The rate at which a new retroviral produces side-effects between cohorts
e. The rate of eventual HIV infection in HIV-negative enrollees

**Correct answer:** B

B is the correct answer because it is the only risk factor in the listed choices which may be obtained from a case-control study. A case-control study specifically collects data about risk factors based on presence or absence of disease. A cohort study collects data on the development of disease based on the presence of absence of risk factors. Answers A, C and E would be feasible in a cohort study. Answer D is an example of an outcome feasible when an intervention is given to the groups to gauge efficacy or side-effects of the intervention. This is specific to clinical trial studies.
The enzyme-linked immunosorbent assay (ELISA) for the HIV virus has a sensitivity and specificity of 99.9%. Therefore, in a population of 100,000 people where the prevalence of HIV is 1%, there will be 1 false negatives and 99 false positives. Likewise, there will be 999 true positives and 98,901 true negatives.

100 HIV+: 999 T+ and 1 T-
99000 HIV-: 98901 T+ and 99 T-

If an analysis was made of a blood donor pool in which blood had already been screened for HIV and now the prevalence of HIV was 0.1%, how would the sensitivity, specificity, positive predictive value (PPV) and negative predictive value (NPV) change?

a. Decreased sensitivity, Decreased specificity, Same PPV, Same NPV
b. Decreased sensitivity, Decreased specificity, Decreased in PPV, Decreased in NPV
c. Same sensitivity, Same specificity, Same PPV, Same NPV
d. Same sensitivity, Same specificity, Decreased PPV, Decreased NPV
e. Same sensitivity, Same specificity, Decreased PPV, Increased NPV

**Correct answer:** E

With any test, no matter its sensitivity and specificity, changing the prevalence of the disease will change the positive and negative predictive values of the test. In this case, as the prevalence decreases, positive predictive value will decrease and negative predictive value will increase.

Taking the example from the question stem, a prevalence of 0.1% would give us about 100 true positives and 100 false positives. This is a positive predictive value of 50% as compared to 91% when the prevalence was 1%. The negative predictive value goes up a fraction of a percent, from 98901/98902 to 99800.1/99800.2.

100 HIV+: 99.9 T+ and 0.1 F-
99900 HIV-: 99.9 F+ and 99800.1 T-

Sensitivity and specificity are inherent features of a test, and do not change if prevalence changes. Therefore, the answer is choice E.
A researcher develops a new test to detect lung cancer at an earlier stage. The test picks up the lung cancer in 85% of patients with lung cancer diagnosed by the gold standard and is negative in 90% of patients without lung cancer. The prevalence of lung cancer in the population of 1000 being studied is 3%. What is the sensitivity of the test?

- a. 32.5%
- b. 66%
- c. 85%
- d. 87.5%
- e. 90%

**Correct answer:** C

Sensitivity is defined as the probability that a test will be positive in individuals who are already known to have the disease, in this case, 85% (choice C).

90% (Choice E) refers to the specificity, or the probability that the test will be negative in individuals who are known to not have the disease in question.
An aspiring medical student opens up his Step 1 score report and is elated to find that his score is 230, a 92 on the 1-100 scale! The score report indicates that the mean on the exam was 215 and the standard deviation is 15. Assuming a normal distribution, in which percentile is the student's score?

a. 63.5%
b. 77.3%
c. 84.1%
d. 97.7%
e. 99.9%

correct answer: C

This question tests your understanding of cumulative percentiles and standard deviations (sigma) in the Normal Distribution. You should know that 1(sigma) = 34.1%, 2(sigma) = 95.4%, 3(sigma) = 99.7%, and of course that 0(sigma) = 50%. Here, the mean is 215 and the deviation = 15 (230-215). This corresponds to .75 (sigma) (deviation / std. deviation). Therefore, the likely answer is C. About 34.7% of the population is included in the .75 std. deviation (in the positive direction), this value is then added to the 50% level of the mean, which is equal to 84.1
A 45-year-old female patient consults her physician with concerns over her risk of dying from cancer. Based on current statistics, her physician tells her that the leading cause of cancer-related deaths in females is due to which of the following malignancies?

- a. Breast
- b. Cervical
- c. Colorectal
- d. Lung
- e. Ovarian

Correct answer: D

Lung cancer is the leading cause of malignancy-related deaths in women, followed by breast cancer (Choice A) and colorectal cancer (Choice C). Ovarian cancer (Choice E) is the fifth leading cause of cancer deaths in women. Cervical cancer (Choice B) is the fourteenth leading cause of cancer deaths in women, and the third leading cause of cancer deaths in women world-wide.

Breast cancer has the highest incidence in women, followed by lung cancer and colorectal cancer.
A 17-year-old male comes to his physician after having unprotected sexual intercourse, concerned that he might have contracted HIV. In counseling this patient about the screening test most widely used, you tell the patient that if the result is positive, a confirmatory test will be needed. This screening test can be said to have a high level of which of the following?

- a. Negative predictive value
- b. Positive predictive value
- c. Prevalence
- d. Sensitivity
- e. Specificity

**Correct answer:** D

The screening test most commonly used to test for HIV is the enzyme immunoassay, otherwise known as the ELISA test. It works by detecting HIV antibodies that may be present in a patient's serum. The ELISA has a high sensitivity, defined as the proportion of positive cases that are correctly identified as such. In contrast, specificity (Choice E) refers to the proportion of negative cases that are correctly identified as such. For any screening test, there is a trade-off between sensitivity and specificity; the higher the sensitivity, the lower the specificity, and vice versa.

Negative predictive value (Choice A) refers to the proportion of patients with negative test results who are correctly diagnosed. Positive predictive value (Choice B) refers to the proportion of patients with positive test results who are correctly diagnosed. Prevalence (Choice C) refers to the total number of cases in a population at a given time, not to be confused with incidence, which refers to the number of cases that develop in a population during a specific time period.
A researcher wants to find out how good a new screening Test A is for detecting disease X. She obtains the results above.

- a. 43/45
- b. 43/100
- c. 98/100
- d. 98/155
- e. 98/200

**correct answer:** D

Choice D (98/155, or true positives over all those with positive tests) is the positive predictive value of the test.

Choice A (43/45, or true negatives out of all those with negative tests) is the negative predictive value.
Choice B (43/100, or true negatives over all those without the disease) is the specificity of the test.
Choice C (98/100, or true positives over all those with the disease) is the sensitivity of the test.
Choice E (98/200) is the proportion of true positives in the sample population.
A randomized double-blind placebo-controlled trial to assess the efficacy of drug X in the treatment of Crohn's disease. 100 people were recruited into the study and were divided equally between the treatment arm and the control group. Over the course of one year, 90 of the 100 people who were treated with drug did not incur side effects, whereas 95 of 100 people in the placebo group did not incur side effects. How many people treated with drug X for one year will suffer from side effects that they would not have otherwise experienced?

a. 5
b. 10
c. 20
d. 25
e. 50

correct answer: C

This question asks you to calculate the Number Needed to Harm (NNH). The NNH is the reciprocal of the Absolute Risk Increase (ARI). That is, NNH = 1/ARI. ARI = absolute value of the difference between incidence in the exposed and incidence in the unexposed. In the case presented, incidence of side effects was the outcome of interest. In the exposed, this incidence was 10 of every 100 people treated with drug X, and 5 out of every 100 people treated with placebo. That means that the incidence of effects solely attributable to drug X in this case is .05, explained by the equation ARI = incidence exposed - incidence unexposed = 0.1-0.05 = 0.05. Thus, NNH = 1/ARI = 1/0.05 = 20.
A physician is trying to determine if exposure to substance X is a risk factor for a disease Y, which has a prevalence of 1 in 10 million. What is the most appropriate study design for her research?

- a. Case-Control
- b. Cross-Sectional
- c. Program
- d. Prospective Cohort
- e. Randomized Controlled

**Correct answer:** A

A Case-Control (Choice A) study, which compares exposures in patients with the outcome of interest and controls, would be the best study design since the outcome is very rare. With an outcome with such a low prevalence, it would be difficult to recruit enough patients to run a prospective cohort study (choice D), which can be used to compare outcomes in two groups with different exposures.

Randomized Controlled Trials (Choice E) and Program Trials (Choice C) are used to compare treatments or interventions. Cross-sectional studies (Choice B) cannot be used to establish risk factors, since all the data is gathered at the same time.
A doctor is developing a new screening test for diabetes. If she wants to minimize the number of false negatives, she should place her blood glucose level cutoff point at which of the above concentrations?

a. Concentration A  
b. Concentration B  
c. Concentration C  
d. Concentration D  
e. Concentration E

**Correct answer:** B

Choice B would maximize the test’s sensitivity and minimize the number of false negatives, people who have the disease but test negative.

Choice A would have a tremendous number of false positives even though it would pick up all the diabetics. Choice C would have a relatively high sensitivity and specificity but would have some false positives and false negatives. Choice D would maximize specificity, and would have no false positives. Choice E would have no false positives but would miss everyone with the disease!
A study reports that a new screening test to detect disease G increases survival of patients from diagnosis to death from 6 months to 1 year. A review of the study finds that the new test does not actually affect mortality from disease G. This error is an example of which kind of bias?

a. Lead time bias  
b. Recall bias  
c. Reporting bias  
d. Sampling bias  
e. Selection bias

**Correct answer:** A

The correct answer is Lead time bias (choice A): the early detection does not lengthen life but only detects it earlier. The space between the two arrow is known as the lead time. Recall bias (Choice B) affects case control studies and studies where patients are asked to remember exposures, where people with and without the disease may remember their exposures differently.

Reporting bias (choice C) refers to a situation in which people with and without a disease may respond differently to an interview or survey. In this case, if the criteria used to report deaths due to disease G differed across study sites, this could bias the results.

Sampling bias (choice D) refers to a sampling of the population that overemphasizes a certain type of group more than it would be emphasized in the general population and thus skews the study results: if this test picked mostly young men with the disease and they have a higher chance of survival than others, this could bias the test results.

Selection bias (Choice E) refers to a study design that looks at a part of the population that ‘stacks the deck’ in favor of certain results: in this case, if the screening test was performed only on people who had no comorbidities and therefore a better overall chance of survival.
**Question**: A medical student seeks to examine the effect of sleep on passing an exam. She gives a survey to her classmates right before the exam to find out whether or not they slept the previous night, and then obtains data on the exam scores, and finds the above results. This project is an example of what kind of study design?

a. Case-Control  
b. Cross-Sectional  
c. Historical Cohort  
d. Prospective Cohort  
e. Randomized Controlled

**Correct Answer**: D

This study is an example of a Prospective Cohort study (choice D) since she investigated the exposure (sleep) and then followed the groups to see who passed and failed (outcome).

A Case Control (Choice A) design would be if she chose students who passed and students who failed (outcome) and then asked them about how much sleep they got the night before the exam (exposure).

A Cross-sectional trial (Choice B) would look at test-scores and sleep habits in the student population but wouldn’t give any information about associations, odds or causality.

A Historical Cohort study design (Choice C) would be similar to a prospective cohort design except that all the events would have already occurred in the past, so the data would all be gathered on events that had already happened.

A randomized controlled trial (Choice E), while problematic ethically, would entail randomized the students into two groups, one of which would get to sleep and the other would not (intervention) and then tracking the test results (outcome).
In assessing the efficacy of natalizumab, a human monoclonal antibody against alpha-4 integrin, in the treatment of multiple sclerosis (MS), a randomized double-blinded placebo-controlled study was performed with a total of 1000 people divided equally between the treatment and placebo groups. The study lasted for three (3) years and during this period, the reduction in relative risk for primary events (relapse of current MS or progression to a more chronic form of MS) was 8%. The absolute risk reduction was smaller because the study group had a low incidence of MS-associated events., 1% in the control group and 3% in the treatment group. How many patients must be treated for three years with natalizumab to prevent a primary event in one person?

a. 2
b. 1
c. 50
d. 100
e. 20

correct answer: C

The question asks you to determine the number needed to treat (NNT) which is the reciprocal of the absolute risk reduction (ARR). That is, NNT = 1/ARR. The absolute risk reduction is the decrease in risk of an event in relation to the control. That is, AR = Incidence in test group - incidence in control group. Here AR = 3% - 1% => 2%. So, NNT = 1/ARR = 1/0.02 = 50. (Don’t forget that you are inverting percentages not numbers!)
55-year-old man visits his primary care physician with a complaint of nodule in the prostate gland. PSP serum test done, PSP of 4 ng/dl is considered normal. The test has a sensitivity of 80% and a specificity of 90%. In a cross-sectional study, 10% of men of this age have prostate cancer. The patient's PSA is 7 ng/dl. What is your best estimate of the likelihood of this man having prostate cancer?

a. 13%

b. 36%

c. 47%

d. 69%

e. 72%

correct answer: C
A 35 year old woman presents with recent onset constitutional symptoms, including daily fevers, a 5 kg weight loss, and fatigue. She also has been “coughing up blood” and has had occasional sinus headaches and runny nose. An initial chest X-ray shows cavitations. A PPD is read as negative. You suspect the patient may have Wegener’s granulomatosis, and want to test her cANCA level. You tell her that in general the probability a positive test result means that she actually has the disease are most dependent on

- The sensitivity of the cANCA assay for Wegener’s
- The specificity of cANCA assay for Wegener’s
- The prevalence of Wegener’s in people with her symptoms
- Her HLA type
- How long she has been symptomatic

**correct answer:** C

The probability that a patient is truly positive for a disease given a positive test result is known as the positive predictive value (PPV). The PPV changes with the prevalence of the disease in a population. When the disease is very prevalent, the PPV will increase; when it is very uncommon, it will decrease. The opposite is true of the negative predictive value.

Sensitivity and specificity of a test are fixed test characteristics that are not affected by disease prevalence.

The other choices are not relevant for this disease and do not answer the question.
A new drug is being developed to increase cardiac contractility in heart failure patients. Assuming the receptor of action of this drug also increases heart rate, which of the following is likely involved in the pathway of this drug's signaling?

- a. Diacyl glycerol
- b. Inositol triphosphate
- c. Phospholipase C
- d. Protein kinase A
- e. Zinc ion

**Correct answer:** D

The drug in development is likely acting on the Beta-1 receptor in cardiac tissue, simultaneously increasing both contractility and heart rate. Beta-1, Beta-2, D1, H2, glucagon, and V2 receptors are Gi-protein-linked which act via adenylcyclase to create cAMP and eventually lead to increased protein kinase A as a second messenger. Alpha-1, M1, M3, H1 and V1 receptors are Gq-protein-linked, and act via phospholipase C to create either IP3 or DAG as second messengers. Zinc ion is a distractor in this question stem.
Some competitive weight-lifters use substances that increase the activity of nitric oxide synthase (NOS) before a workout to increase the blood-supply to muscles. Which of the following is a plausible mechanism by which one of these substances could work?

a. Decrease in intracellular cGMP
b. Increased in intracellular calcium
c. Decreased deamination of arginine
d. Inhibition of myosin light chain phosphatase
e. Increased oxidation of NADPH

**Correct answer:** C

The question asks not for the mechanism of NOS, which involves an increase in intracellular cGMP (distorted in answer choice A) that leads to decreased intracellular calcium concentrations (distorted in answer choice B) and stimulation of cGMP-dependent myosin light-chain phosphatase (distorted in answer choice D). Rather, the question asks for ways in which the production of NO (think prior to its activity) could be increased, that is for ways to make NOS more efficient. The production of NO begins with the addition of NADPH and oxygen to arginine. Decreased deamination of arginine (answer choice C) would increase the amount available to produce NO. Increased oxidation of NADPH to NADP would decrease the amount of a cofactor necessary to convert arginine to NO (answer choice E).
A girl who is deaf and mute dies immediately after her teacher yells at her during a lecture in class. The parents report that her brother was also deaf and mute and died suddenly during a fight. Concerned about their third child, they consult a pediatrician who during a battery of tests notices an elongated QRS complex. Upon referral to a pediatric geneticist, it is determined that the child has a loss-of-function mutation on chromosome 11p15.5 in the gene LQT1, a subunit of the slow delayed rectifier potassium channel. How does this mutation give rise to the ECG finding?

- Increased phase 1 of ventricular action potential
- Decreased phase 2 of ventricular action potential
- Increased phase 3 of ventricular action potential
- Decreased phase 0 of atrial action potential
- Increased plateau of atrial action potential

**Correct answer:** C

The key to this question is to recognize the function of a delayed inward rectifier channel. Inward rectifier channels pass positive current more easily into cells than outside. That is their iV curves display at least one discontinuity looking like "hockey sticks".

Inward rectifying potassium channel will favor the net movement of potassium into the cell causing a repolarization. Loss of function of this channel would cause a decrease in repolarizing current increasing the time that the ventricular myocyte needs to return to baseline. Since the elongation is in the QRS complex, the dysfunction is occurring in ventricular repolarization so disregard answer choices D and E. Phase 1 is not found in ventricular action potentials. Phase 2 is the depolarization of the myocyte so a decrease in its duration would mean an decrease in QRS duration, not an increase.

I AM NOT SURE OF THIS AS A DELAYED RECTIFIER IS NOT AN INWARD RECTIFIER. DELAYED RECTIFIER ACTS IN THE PLATEAU OF THE ACTION POTENTIAL (USUALLY), WHEREAS INWARD RECTIFIER MAINTAIN RESTING POTENTIAL WHEN HYPERPOLARISATION OCCURS.
A 56-year-old man complains of increasing epigastric pain and diarrhea of one month's duration. He reports one episode of vomiting bright red blood. Endoscopy reveals multiple small ulcerations throughout the stomach and duodenum. A secretin stimulation test demonstrates elevated gastrin levels in response to secretin. Pharmacologic inhibition of which cell type would result in symptomatic relief in this patient?

a. Chief cell  
b. G cell  
c. I cell  
d. Parietal cell  
e. S cell

**correct answer: D**

This man likely suffers from Zollinger-Ellison syndrome, a disorder where gastrin levels are ectopically increased, resulting in higher levels of HCl production in the stomach. The source of excess gastrin is often a gastrinoma in either the duodenum or pancreas. Gastrin stimulates parietal cells (D) in the stomach to produce HCl, resulting in ulceration of the stomach and small bowel. Therefore, use of proton pump inhibitors or H2 blockers can reduce acid output by the parietal cells, and improve symptoms.

Chief cells (choice A) are also located in the stomach, but produce pepsin for protein digestion.

G cells (choice B) are found in the antrum of the stomach. They produce gastrin, but are not responsible for the abnormally high levels found in Zollinger-Ellison syndrome.

I cells (choice C) are found in the duodenum and jejunum, and they produce cholecystokinin, which increases pancreatic secretion and gallbladder contraction, and decrease gastric emptying.

S cells (choice E), found in the duodenum, produce secretin, which increases pancreatic bicarbonate secretion and decreases gastric acid secretion.
A 45 year old woman with a history of 30 lbs weight loss and fatigue over the last 4 months presents with complaints of coughing up blood-tinged sputum and chest pain worsened when she "breathes in". She reports having had pain in the left thigh, which later “moved” to her right leg, and now has pain in her right leg. She has an elevated D-dimer. Physical examination is significant for scleral icterus and right upper quadrant tenderness. Which of the following molecular markers is most likely to yield this patient's diagnosis?

- a. AFP
- b. CEA
- c. CA-125
- d. CA 19-9
- e. beta HCG

**correct answer: D**

This patient clinical picture is consistent with a pulmonary embolism, secondary to migratory thrombophlebitis (also called "thrombophlebitis migrans", meaning that the thrombi are moving from vein to vein, which leads to pain associated with the thrombi moving as well). Malignancy predisposes to hypercoagulability, and venous thrombosis in the setting of neoplasms is called Trousseau syndrome. However migratory thrombophlebitis (or thrombophlebitis migrans) is seen almost only in pancreatic and lung cancers. CA 19-9 is found in up to 93% of pancreatic cancers.

AFP is found in liver cancers. The RUQ tenderness is most likely due to the tumor located in the pancreatic head, obstructing biliary flow and back up in the common bile duct. It is not known to cause migratory thrombophlebitis.

CEA is found in colon cancer, which is not associated with migratory thrombophlebitis.

Beta HCG is associated with germ cell tumors, such as ovarian cancer, but is uncommonly associated with migratory thrombophlebitis.
Familial Adenomatous Polyposis (FAP) is a form of colon cancer with mutations in the following pathway:

Wnt ----| APC -----| beta-Catenin ----> increased transcription.

In this pathway, the APC protein is considered to be a:

a. Carcinogen  
b. Proto-oncogene  
c. Transcription Factor  
d. Transmembrane Protein  
e. Tumor Suppressor

**Correct answer:** E

Note that mutations in APC cause FAP. It is considered to be the "gatekeeper" in this pathway. As with most tumor suppressors, mutations that cause cancer typically are recessive and require two mutated copies. If one gene is already inactivated (for instance, via inheritance of a bad copy), a mutation in the other gene can result in a Loss of Heterozygosity and lead to dysregulated signaling in the Wnt/APC/beta Catentin pathway.
A 22 year old female patient presents with shortness of breath, worsened by smoking. Transbronchial biopsy reveals cells from the monocyte lineage. On electron microscopy, structures are seen that look like “tennis rackets”. A chest X-ray reveals a honey combing appearance of the lungs. Her WBC is 7,000, and she has a body temperature of 37.1. Which is the most likely diagnosis?

a. Bacterial pneumonia  
b. Viral pneumonia  
c. Langerhans cell histiocytosis  
d. Chronic asthma  
e. Chronic obstructive pulmonary disease

**Correct answer:** C

This patient has Langerhans cell histiocytosis, an interstitial lung disease that is epidemiologically related to tobacco smoking. It chiefly affects young adults, primarily occurring in the third or fourth decades of life. LCH is histologically characterized by abnormal infiltration of the lungs by activated Langerhans cells. Langerhans cells are differentiated cells of the dendritic cell system and are closely related to the monocyte-macrophage line. They may be identified by immunohistochemical staining or by the presence of Birbeck granules via electron microscopy. These antigen-presenting cells are normally found in the skin, reticuloendothelial system, heart, pleura, and lungs. The accumulation found in the lungs is hypothesized to occur in response to exposure to cigarette smoke. The granulomatous infiltrates seen in PLCH are composed of Langerhans cells, eosinophils, lymphocytes, macrophages, plasma cells, and fibroblasts, which form nodules centered on the terminal and respiratory bronchioles, causing destruction of the airway walls.

Answer A is incorrect; bacterial pneumonia would likely to present with fever and leukocytosis, which this patient does not have.

Answer B is incorrect; the picture is not consistent with a viral pneumonia, which would show diffuse infiltrate in the lungs or a normal X-ray. Honey combing suggests fibrosis, a more chronic process involving destruction and remodeling of airway walls.

Answer D is incorrect; the patient could have asthma, but the Birbeck granules point out more specifically to LCH.

Answer E is incorrect; the patient could have COPD, but the finding of Birbeck granules, as well as the young age, are more suggestive of LCH.
A 15 year old male patient presents with a chronic weakness for many months. Laboratory workup reveals a hematocrit of 20%, a decreased mean corpuscular volume, and an increased RDW (red blood cell distribution width). There is no history of metal poisoning or substance abuse. The patient denies taking any medications. A bone marrow aspirate revealed erythroid hyperplasia. Biochemical assays reveal absence of a cofactor needed for synthesis of a key enzyme. That cofactor is:

- Riboflavin
- Biotin
- Thiamine
- Pyridoxine
- Cobalamin

**Correct answer:** D

This patient has sideroblastic anemia. The figure illustrates sideroblasts, which are nucleated (immature) erythrocytes with granules of iron in their cytoplasm. In this condition, heme molecules synthesis is abnormal in the mitochondrion, which is one of the organelles participating in making heme. Abnormal heme leads to deposits of iron in the mitochondria that form a ring around the nucleus of the developing red blood cell. The enzyme delta aminolevulinic acid (ALA) is essential in the biosynthesis of heme. Delta ALA requires pyridoxine (vitamin B6) and copper as cofactors.

- Answer A is incorrect; riboflavin is a cofactor used by FADH2
- Answer B is incorrect; biotin is used in carboxylation reactions
- Answer C is incorrect; thiamine is a cofactor used in the TCA cycle and the pentose phosphate pathway
- Answer E is incorrect; cobalamin is involved in transfer of methyl groups
A 9-month-old baby is brought to her pediatrician because she has suffered recurrent respiratory tract infections. At birth, she had coarse facial features and musculoskeletal deformities. Microscopic examination of her cells reveals large inclusion bodies due to the accumulation of undegraded products in lysosomes. An enzyme deficiency in which organelle is responsible for her disease?

a. Endoplasmic reticulum  
b. Golgi apparatus  
c. Mitochondria  
d. Nucleus  
e. Peroxisome

**Correct answer:** B

This patient has the clinical presentation of I cell disease, or mucolipidosis II. The disease is caused by a deficiency in N-Ac-glucosamine phosphotransferase in the Golgi apparatus (choice B). This enzyme is responsible for converting specific mannose residues to mannose-6-phosphate in glycoproteins (i.e. hydrolases) destined for lysosomes. Without the enzyme, these glycoproteins follow the default pathway of constitutive exocytosis from the cell. Therefore, degradative enzymes are missing from lysosomes, and undegraded products accumulate to form the characteristic intracellular inclusion bodies.

The other answer choices are not involved in I cell disease.
A one-year-old boy is brought to you because his mother complains he has not said any words yet. On further history, she reveals that he does not babble, and is unable to sit unaided or orient to voice call when his name is called. Physical exam is remarkable for a large forehead, long face with a high-arched palate. Which of the following mechanisms is likely responsible for this child's condition?

a. Autosomal dominant inheritance  
b. Autosomal recessive inheritance  
c. DNA point mutation  
d. Mitochondrial inheritance  
e. X-linked trinucleotide expansion

**Correct answer:** E

This child likely has fragile X syndrome. The failure to attain pediatric milestones of a normal one-year-old child is common considering fragile X is the most common cause of mental retardation in males. It is transferred in an X-linked manner due to a progressive expansion of CGG repeats in the FMR1 gene region. This leads to a failure in the production of an RNA-binding promoter and results in the classic fragile X facies, large testes and mental retardation.
The Hedgehog pathway is one of the most commonly affected signaling pathways in Basal Cell Carcinoma. The diagram of the pathway is as follows:

\[ \text{Hedgehog (Hh)} \rightarrow \text{Patched (PTCH)} \rightarrow \text{Smoothened (SMO)} \rightarrow \text{GLI}. \]

In this case, PTCH normally suppresses SMO, thus preventing increased signaling and gene transcription. In this sense it is acting as a tumor suppressor.

**Correct answer:** D

In this case, Patched (PTCH) normally suppresses SMO, thus preventing increased signaling and gene transcription. In this sense it is acting as a tumor suppressor.
Which band on the above image predominantly contains microfilaments?

a. A band  
b. H band  
c. I band  
d. M line  
e. Z line

**correct answer:** C

Muscle contraction is an important concept to have down. Check this link for a great example:
http://www.lab.anhb.uwa.edu.au/mb140/CorePages/Muscle/Images/Mus1ani.gif  
Actin, a microfilament, is the main component of the I band. H, I, and Z are the only components that SHRINK during muscle contraction. Be ready for a question on function.
A 72-year-old man presents with large, tense, pruritic bullae on the lower abdomen, groin, and inner thighs. Skin biopsy reveals IgG antibodies with linear immunofluorescence at the epidermal basement membrane. To what cell junctions are the antibodies binding?

- a. Adherens junctions
- b. Desmosomes
- c. Gap junctions
- d. Hemidesmosomes
- e. Tight junctions

**Correct answer:** D

This man's clinical presentation suggests bullous pemphigoid, which is an autoimmune disorder with IgG antibody against epidermal basement membrane hemidesmosomes (choice D). This disorder is similar, but not as severe as pemphigus vulgaris, which affects desmosomes (choice B). Pemphigus vulgaris usually affects the oral mucosa as well, while bullous pemphigoid mostly spares the oral mucosa.

Adherens junctions (choice A), gap junctions (choice C), and tight junctions (choice E) are other cell junctions that are not involved in bullous pemphigoid.
A 37 year-old woman comes to the physician with multiple symmetric arthralgias with considerable morning stiffness. She also experienced difficulty swallowing and shortness of breath upon exertion. Her hands show significant swelling with red patches over her knuckles with a sausage-like appearance. She has a mild malar rash on her face and slight discoloration of her eyelids. Chest X-ray shows diffuse interstitial infiltrates. There are no joint deformities noted. Which antibody titers will be highest in this patient?

a. c-ANCA  
b. p-ANCA  
c. Scl-70  
d. RNP  
e. dsDNA

**Correct answer:** D  
D IS THE CORRECT ANSWER:
This question requires that you understand the presentation of mixed connective tissue disease. The patient has multiple symmetric arthralgias with morning stiffness suggesting a rheumatoid arthritis; difficulty swallowing and shortness of breath upon exertion suggesting scleroderma; red patches over her knuckles suggesting dermatomyositis; a sausage-like appearance suggesting spondyloarthritis; mild malar rash on her face and slight discoloration of her eyelids suggesting systemic lupus erythromatosus. This has now been considered as a separate disease entity since it has a distinct autoantibody pattern to the ribonucleoprotein (anti-RNP).

A IS INCORRECT: Wegener’s granulomatosis is associated with antibodies to c-ANCA.

B IS INCORRECT: Microscopic polyarteritis is associated with antibodies to p-ANCA.

C IS INCORRECT: Systemic scleroderma is associated with antibodies to Scl-70.

E IS INCORRECT: Systemic Lupus Erythromatosus is associated with anti-dsDNA.
A 38 year-old woman presents with shortness of breath and cough productive of blood-tinged sputum. Patient has been unresponsive to antibiotic therapy but showed some improvement on corticosteroid therapy. She is febrile, blood pressure 124/82 mmHg, pulse 74 and respiratory rate of 19/min. Laboratory studies reveal an erythrocyte sedimentation rate (ESR) of 90/min, anemia, and circulating anti-neutrophil cytoplasmic antibodies of c-ANCA type. Urinalysis indicates dysmorphic red cells and red cell casts.

Which of the following antibodies is most likely to be found in the patient's serum?

a. Anti-Proteinase-3
b. Anti-Myeloperoxidase
c. Anti-Centromere
d. Anti-dsDNA
e. Anti-mitochondria

**Correct answer:** A

A IS THE CORRECT ANSWER:

This question presents a classic presentation of Wegener’s granulomatosis. This disease has lung (hemoptyis, nasal mucosal ulceration, saddle-nose deformity) and renal (rapid progressive glomerulonephritis, hematuria) manifestation. Lung involvement that does not resolve with antibiotic therapy should raise a suspicious of a non-infectious etiology. To choose the right option you must recall that an anti-cANCA is associated with Wegener’s and antibodies are directed specifically against the anti-proteinase 3.

B IS INCORRECT: Anti-myeloperoxidase is associated with microscopic polyangiitis. Presentation is similar to Wegener’s but without granulomas.

C IS INCORRECT: Anti-centromere is associates with limited scleroderma with a classic presentation of CREST (Calcinosis, Reynauld’s phenomenon, Esophageal dysmotility, Sclerodactyly and Telangetasia)

D IS INCORRECT: Anti-dsDNA is associated with systemic lupus erythematosus.

E IS INCORRECT: Anti-mitochondria is associated with primary biliary sclerosis.
A 27 year-old man presents with numerous erosions of his mucous membranes. Examination of his oral cavity reveals painful oral ulcers and a friable mucosa. Immunofluorescence evaluation demonstrates intraepidermal band of IgG. A biopsy of the lesion shows breakdown of intercellular epithelial connections. Which of the following is the most likely diagnosis in this patient?

a. Herpetic ulcers
b. Measles
c. Dermatitis herpertiformis
d. Pemphigus vulgaris
e. Bullous pemphigoid

**Correct answer:** D

**D is the correct answer:**
This question requires that you differentiate between intraepidermal IgG as in pemphigus vulgaris and deeper layers of the skin involving the dermal-epidermal junction as in Bullous pemphigoid. The latter produces tense blisters that do not usually ulcerate. Furthermore, oral involvement typically occurs in pemphigus vulgaris.

A is incorrect: There is no intraepidermal IgG deposition in herpertic ulcers.

B is incorrect: There is no intraepidermal IgG deposition in Measles.

C is incorrect: Dermatitis herpertiformis is an extraintestinal manifestation of celiac’s disease. This rash is characterized by IgA and C3 deposition, not IgG.

E is incorrect: same reasons describe in choice D.
A 44 year old female presents with reddened, inflamed patches with a flaky silver-white layer on her left knee. You, as the physician, diagnosed the condition with psoriasis. What is the underlying cause of this condition?

a. A defect in the keratinocyte intermediate filaments
b. An increase in mitotic activity of the cells in the malpighian layer of the epidermis
c. Exposure of unprotected skin to UV light
d. Squamous cell carcinoma arising from cells of the stratum spinosum
e. None of the above

**Correct answer:** B

Psoriasis is a skin condition in which increase in mitotic activity of the cells in the malpighian layer of the epidermis that have shorter than normal cell cycle. Redness is due to an increase of blood flow to nourish the accelerated growth.

(A) suggests Epidermolysis bullosa.
(C) is indicative of skin damage due to radiation.
(D) Skin cancer has no association with psoriasis.
A 37-year-old man has multiple blisters located in his mouth. He complains that they are painful, burst easily and make it difficult to eat. A biopsy of one of the blisters was stained and shown above. Immunofluorescent antibodies directed against which protein would produce a similar pattern to the biopsy?

a. Cadherin  
b. Fibronectin  
c. Integrin  
d. Keratin  
e. Laminin

**Correct answer: A**

This patient is suffering from Pemphigus Vulgaris, as demonstrated by the clinical presentation (multiple, flaccid, painful blisters) and Immunofluorescence biopsy. Note the intercellular IgG deposits in the epidermis and the early intraepidermal vesicle caused by acantholysis.

In Pemphigus Vulgaris, autoantibodies are directed against the desmosomes which attach neighboring cells to each other. The questions asks you to take this information one step further, and recall from your Cell Biology that Desmosomes (macula adherens) are members of the Cadherin (Choice A) family of cell adhesion molecules.

Fibronectin (Choice B) is a ECM glycoprotein that binds to integrins, collagen, fibrin and heparan sulfate. It would produce a different staining pattern and is not involved in Pemphigus.

Integrins (Choice C) are receptors that mediate attachment between a cell and the tissues or ECM surrounding it. They also play a role in cell signaling and thereby define cellular shape, mobility, and regulate the cell cycle. They are not involved in Pemphigus and would produce a different staining pattern.

Mutations in the gene for Keratin (Choice D) lead to Epidermolysis bullosa, a disorder characterized by recurrent blisters. Autoantibodies are not involved in the pathogenesis of this disorder, and immunofluorescence staining to keratin would produce a different pattern.

Laminin (Choice E) is a protein found in the extracellular matrix that forms the basement membrane. It is not involved in Pemphigus and would produce a different staining pattern.
A 2-day-old infant is brought to your office by his mother, who says she has been noticing copious amounts of blood in his stool for the past 2 days. The patient is tachycardic but has not been experiencing any pain. What embryonic structure is likely to be responsible for this patient’s condition?

- a. Ductus Arteriosus
- b. Ductus Venosus
- c. Umbilical Artery
- d. Urachus
- e. Vitelline Duct

**Correct answer:** E

This patient likely has a lower GI bleed from a Meckel’s Diverticulum. This true diverticulum is a remnant of the vitelline duct and often contains ectopic gastric mucosa which can cause bleeding and perforation. The bleeding is often painless.

The ductus venosus is a shunt that bypasses the liver and carries blood from the umbilical vein directly to the IVC. Its remnant is the ligamentum venosum.

The ductus arteriosus is a shunt that bypasses the lungs to carry blood from the pulmonary artery to the aortic arch. Its remnant is the ligamentum arteriosum.

The urachus becomes the median umbilical ligament.

The 2 umbilical arteries becomes the medial umbilical ligaments.

The vitelline duct is a connection with the yolk stalk and bowel, but normally obliterates during week 7 of development.
The greater omentum is derived from which of the following embryonic structures?

a. Dorsal mesoduodenum
b. Dorsal mesogastrium
c. Pericardioperitoneal canal
d. Pleuropericardial membranes
e. Ventral mesentery

**correct answer:** B

The correct answer is B. Both the omental bursa and the greater omentum are derived from the dorsal mesogastrium, which is the mesentery of the stomach region.
The dorsal mesoduodenum (choice A) is the mesentery of the developing duodenum, which later disappears so that the duodenum and pancreas come to lie retroperitoneally.
The pericardioperitoneal canal (choice C) embryologically connects the thoracic and peritoneal canals.
The pleuropericardial membranes (choice D) become the pericardium and contribute to the diaphragm.
The ventral mesentery (choice E) forms the falciform ligament, ligamentum teres, and lesser omentum.
A 15 year-old boy presents to your office complaining of a painful mass on the right side of his neck for one week. Last week he had a sore throat and productive, non-bloody cough that has since gone away. He has noticed that the mass usually becomes painful whenever he is sick with a sore throat, but it is never completely gone. He appears anxious because he cannot remember the last time the mass was not present. On physical exam, the mass is immobile and slightly painful to touch and his oropharynx exam is clear. What is the likely etiology of this patient's mass?

a. Bacterial abscess
b. Ectopic thyroid tissue
c. Incomplete involution of branchial cleft one
d. Incomplete involution of branchial cleft three
e. Neoplasm

**Correct answer:** D

This patient's complaint is likely due to a branchial cleft cyst, due to an incomplete involution of branchial clefts two, three or four during the 7th week of embryonic development. Because they are lined with ectoderm, incomplete involution leaves behind an entrapped epithelium-lined cyst that may persist into adulthood, existing as a painless lateral neck mass only painful during infection. Answer choice A, bacterial abscess, would present with a more dire physical exam, perhaps draining pus, and a patient with febrile illness. Answer choice B, ectopic thyroid tissue, may be found at the base of the tongue or as part of a thyroglossal duct cyst, both of which are ruled out by normal physical exam. Incomplete involution of branchial cleft one, answer choice C, would lead to a disruption in the external auditory meatus, the normal derivative of cleft one, but would not result in a cyst. Neoplasm of the lymph nodes, answer choice E, is unlikely given the chronic nature of the cyst and the lack of other symptoms of lymphoma.
A 10-year-old boy complains of increasing weakness in his legs. His mother notes that the patient is no longer as active as he once was and no longer can participate in gym class due to fatigue. On physical exam, it is noted that the patient uses his hands to help him raise himself off the floor, first getting on his knees then walking his hands back to his legs. His blood tests are notable for an elevate CPK. Which protein is most likely decreased in this patient?

a. Actin  
b. Alpha-1 Antitrypsin  
c. Collagen type I  
d. Dystrophin  
e. Fibrillin

**Correct answer:** D

The patient is suffering from Duchenne muscular dystrophy which is caused by a mutation of the dystrophin (option D) gene on the X chromosome. It is X-linked recessive so it mainly seen in males. The elevated CPK is a sign that there is muscle breakdown which is the reason for the fatigue. The description of the patient raising himself is called the Gowers' Sign.

A deficiency of alpha-1 antitrypsin (option B) is the caused of liver failure and early-onset panacinar emphysema.

A deficiency in type I collagen (option C) is seen in osteogenesis imperfecta.

A mutation in the fibrillin gene is the cause of Marfan syndrome notable for tall patients with severe nearsightedness, lens subluxation, arachnodactyly, hyperextensible joints, and risk of aortic dissection. It is thought to be due to unopposed action of TGF-beta (since fibrillin inhibits TGF-beta in normal patients).

Actin is found in all eukaryotic organisms and primary building blocks in microfilaments and thin filaments (in sarcomere attach to Z line).
A patient presenting to your office has mildly short stature, small epiphyses in the long bones, and early onset osteoarthritis. The patient has multiple epiphyseal dysplasia, a disease of type IX collagen. Which of the following processes is likely have to been impaired in this patient?

a. Endochondral ossification  
b. Intramembranous ossification  
c. Osteoblast differentiation  
d. Osteoclast differentiation  
e. Type I collagen formation

**Correct answer:** A

You need not know anything about this disease to answer this question correctly. The answer is A, endochondral ossification, because this describes the process of ossifying the hyaline cartilage that originally exists in the epiphyses of long bones when a child is first growing. This region was described as being small (or dysplastic) in the question stem, and it can logically be inferred from the question stem that this process of normal development has been impaired in this patient.

B. Intramembranous ossification describes the formation of bone directly from mesenchyme that has formed a membranous sheath. There is no cartilage step in this type of bone formation. This type of bone formation occurs in the bones of the skull, not in the long bones of the limbs.

C, D. Both osteoblasts and osteoclasts are involved in bone remodeling. In the adult, osteoblast activity (laying down of new bone) and osteoclast activity (digestion of old bone) should more or less balance each other out. When there is more osteoclast activity than osteoblast activity, osteoporosis (“porous bone”, decreased bone mass and density) occurs. Osteoblasts are derived from mesenchymal cells (similar to fibroblasts in the skin and other connective tissues). Osteoclasts are of hematopoietic origin similar to that of macrophages and monocytes. Defects in the differentiation of either of these cell lines would result in an imbalance of bone reabsorption and formation, but would not be specific to just the epiphyses of long bones as described in the case.

E. Type I collagen is found in bone, but defects in its formation would manifest as brittle bones throughout all bones in the body. Defects in type I collagen formation results in osteogenesis imperfecta.

Also note that the specific protein defect described, type IX collagen is found in hyaline cartilage and plays a role in stabilizing the more abundant type II collagen that is also found there. Hyaline cartilage is the type of cartilage found in the epiphyseal growth plates before ossification.


Moore and Persuad.  The Developing Human:  Clinical Oriented Embryology, 8th Ed.  pp.339-343

An infant is born with a nasolacrimal duct that has failed to develop a lumen. Between which two of facial prominences does the nasolacrimal duct develop?

- a. Lateral nasal and mandibular
- b. Lateral nasal and maxillary
- c. Maxillary and mandibular
- d. Medial nasal and maxillary
- e. Right and left medial nasal

**Correct answer:** B

Option B (lateral nasal and maxillary) is correct. The nasolacrimal duct develops from an ectodermal thickening in the floor of the nasolacrimal groove between the lateral nasal and maxillary prominences. The epithelial cord normally canalizes via cell degeneration between the lacrimal sac and inferior nasal meatus, but this degeneration may fail to occur.
A male infant has a cleft lip on one side that does not involve the alveolar process or the hard palate. A unilateral cleft lip results from the failure of which two facial prominences to merge?

a. Lateral nasal and maxillary
b. Left and right medial nasal
c. Maxillary and mandibular
d. Medial nasal and maxillary
e. Lateral nasal and Medial nasal

**Correct answer:** D
Choice D (medial nasal and maxillary) is correct. The upper lip is formed when the two medial nasal prominences merge with the two maxillary prominences. A unilateral cleft lip results from failure of the union of the medial nasal and maxillary prominences on one side. If the failure is complete, a cleft of the maxillary alveolar process (between the canine and lateral incisor teeth) and a left of the primary palate also are present. Most cleft lips occur in males, whereas cleft palate is more common in females.

Choice A (lateral nasal and maxillary) fusion of these structures forms the lacrimal duct.
An infant is born to a healthy mother. The infant has some immediately noticeable abnormalities. The infant shows a small mandible, and other facial abnormalities. You inform the mother that her infant has Treacher Collins syndrome. The abnormalities of this symptom is due to:

a. A defect in neural crest cells from the first branchial arch
b. A defect in mesodermal cells from the first branchial arch
c. A defect in mesodermal cells and neural crest cells from the second branchial arch
d. An excessive consumption of ethanol during pregnancy
e. A teratogen such as isotretinoin or other Vitamin A derivative

correct answer: A
A- Treacher Collins syndrome is due to a fail of neural crest cell migration in the first branchial arch.

B- Incorrect. The defects seen in this infant are due to neural crest cells not mesodermal cells. Otherwise this answer would be correct.

C- Incorrect. The second branchial arch is responsible for the Stapes, Styloid process, lesser horn of the hyoid, and Stylohyoid ligament, NOT the mandible.

D- Describes Fetal alcohol syndrome

E- Retinoids have several defects, especially of the skin, but usually NOT mandibular hypoplasia.
A 4-year old child is found to have hypotonia, hyporeflexia, and weakness in his left lower limb, with varying amount of atrophy. His parents also described some degree of incontinence. On examination, he is found to have a spinal bifida. Which of the following nutrients was likely to be deficient in the mother's diet during pregnancy?

- a. Vitamin B1 (thiamine)
- b. Vitamin B12 (cobalmin)
- c. Folate
- d. Vitamin C
- e. Vitamin B6 (pyridoxine)

**Correct answer:** C

Low maternal intake of folate is associated with development of spinal bifida. The other choices do not characteristically result in spinal bifida.
A 30-year-old pregnant woman G3P2002 in her 22nd week of gestation is being counseled by her obstetrician after her fetus has been diagnosed with bilateral renal agenesis by ultrasound. Which of the following is consistent with this diagnosis?

- a. Neonates with this finding generally have no complications
- b. Neonates with this finding usually have Potter’s syndrome
- c. This disease often occurs when the allantois persists
- d. This finding is associated with polyhydramnios during pregnancy
- e. This is a common finding on prenatal ultrasound

**Correct answer:** B

Failure in ureteric bud development results in renal agenesis. Bilateral renal agenesis is associated with Potter’s syndrome (choice B), which is a constellation of deformed limbs, wrinkly skin, and abnormal facial appearance.

Oligohydramnios instead of polyhydramnios (choice D) is seen in bilateral renal agenesis. Much of the amniotic fluid volume comes from fetal urine production which would be severely lacking in bilateral renal agenesis.

Unilateral renal agenesis is a more common finding on prenatal ultrasound (choice E). Bilateral renal agenesis is rare.

Bilateral renal agenesis is not compatible with life (choice A) secondary to many complications unless a kidney transplant with a suitable donor is performed.

Persistence of the allantois (choice C) would result in an urachal fistula, forming a direct connection between the outside of the body and the urinary bladder.
A 23 year old man is hospitalized for shaking chills, flank pain, and painful urination. He reports that the pain first started with urination last week then developed into flank pain 3 days ago. He then decided to seek medical attention because of new onset shaking chills. Physical exam shows temperature 38.2°C, BP 90/50, pulse 125bpm and severe costo-vertebral angle tenderness. Urinalysis show pyuria, bacteriuria, and hematuria. Despite appropriate management, the patient expires. Autopsy is significant for this finding demonstrated in the picture. Which of the following is most consistent with the pathology found at autopsy?

a. Chromosomal analysis in this patient would show a deletion of the WT1 gene  
b. Further urinalysis would reveal elevated vanillylmandelic acid (VMA) levels  
c. Most patients with this finding are asymptomatic  
d. Other autopsy findings may include cysts in the liver  
e. This patient likely has diabetes and hypertension as well

**Correct answer:** C

The image depicts a finding of horseshoe kidney. Horseshoe kidney is a result of a fusion between the inferior poles (90% of the time) of both kidneys. It is the most common fusion anomaly of the kidney. Although patients with horseshoe kidney are at increased risk of urinary tract infections and pyelonephritis, the majority of patients are asymptomatic (choice C) and are diagnosed by coincidence (routine antenatal ultrasonography). This patient unfortunately expired from septic shock secondary to severe pyelonephritis. This pathology is not associated with diabetes and hypertension (choice E).

Cysts in the liver (choice D) may be an additional finding in patients with polycystic kidney disease.

Deletion of the WT1 gene (choice A) is most likely found in patients with Wilm’s tumor.

Elevated VMA levels (choice B) can be found in patients with a pheochromocytoma and in patients with a neuroblastoma.
A 37-year-old woman presents to your office after a positive home-pregnancy test. Because of her advanced maternal age, she is concerned about the possibility of chromosomal abnormalities. Her serum alpha-fetoprotein level result comes back abnormally low for her gestational age. An amniocentesis is performed, as well as the pictured fetal karyotype. Which of the following terms best describes this picture?

- a. Diploidy
- b. Monosomy
- c. Polyploidy
- d. Triploidy
- e. Trisomy

**Correct answer:** E

Trisomy (Choice E) is a form of aneuploidy characterized by the presence of an additional chromosome of a particular type in an otherwise diploid cell (2n+1). Down’s syndrome, which is associated with advanced maternal age and a low maternal serum alpha-fetoprotein level, is the most common type of trisomy (in a live birth, most common trisomy overall would be trisomy 16). In contrast, monosomy (Choice B) is a form of aneuploidy characterized by the loss of one chromosome of a particular type from an otherwise diploid cell (2n-1). An example of monosomy is Turner’s syndrome, in which affected females have a karyotype of 45,XO. Polyploidy (Choice C) is the addition of extra haploid sets of chromosomes to the normal diploid set. Triploidy (Choice D) is an example in which cells contain 69 chromosomes (3n). Diploidy (Choice A) is the normal condition in which cells contain 46 chromosomes (2n).
a 29 yr old woman gives birth to a baby girl. pathologist examines placenta after birth, which of the placental part is derived from mother rather than from fetus?

a. lacunar network  
b. chorion  
c. cytotrophoblast  
d. primary vilus  
e. syncytiotrophoblast

**correct answer:** A

Placental parts derived from mother-cytotrophoblast and syncytiotrophoblast. Both are components of chorion. Primary vilus are processes of chorion that maximize contact with maternal blood. Fetal part-lacunar network that contact with maternal spiral arterioles.
which of the following serve as a stem cell throughout adult life?

a. primary spermatocyte
b. primordial germ cell
c. type A spermatogonia
d. type B SPERMAOGONIA
e. PRIMARY OOCYTE

**correct answer:** C

primordial germ cells arrive in the indifferent gonad at week 4 and remain dormant until puberty. when a boy reaches puberty, primordial germ cells differentiate into type A SPERMATOGONIA WHICH SERVE AS A STEM CELL THROUGHOUT ADULT LIFE....
A 30 year old man is examined after having an episode of angina pectoris during a soccer match. Physical examination reveals a lump above the Achilles tendon and yellowish patches above the eyelids, but is otherwise unremarkable. His BMI is 23, eats a healthy diet and follows a regular exercise routine. His blood work reveals markedly elevated levels of LDL. HDL and triglycerides are within normal limits. Which of the following genetic conditions the patient is most likely to have?

a. Homozygous for ApoB mutation
b. Heterozygous for LDL Receptor mutation
c. Heterozygous for RET mutation
d. Homozygous LDL Receptor mutation
e. Heterozygous for ApoB mutation

**correct answer:** B

Familial hypercholesterolemia (FH) is a common genetic disorder leading to elevated levels of LDL in blood. This may lead to development of atherosclerosis at an early age contributing to coronary artery disease. Physical exam findings include cholesterol deposits on tendons (xanthomas), above the eyelids (xanthelasma palpebrarum), and the outer margins of the iris (arcus senilis corneae).

Individuals with FH most commonly are heterozygous for an LDL Receptor mutation (1 in 500), and heterozygous for an ApoB mutation less often (1 in 1000) (option E). Homozygous individuals with either mutation (Options A and D) are rare and present with a more severe form of the disease in childhood. LDLRAP1 mutations are another cause of the disease but are extremely rare. RET proto-oncogene mutations are seen in MEN-2B, and are not associated with hypercholesterolemia.
A patient presents with hypertension of 160/100, but very weak pulses were felt on palpation of the lower extremities. A radial-femoral delay was observed. Which of the following syndromes is this associated with?

- a. Down's Syndrome
- b. Adult Polycystic Kidney Disease
- c. Kleinfelter's Syndrome
- d. Turner's Syndrome
- e. Marfan's Syndrome

**Correct answer:** D

Patient has coarctation of the aorta, in which arterial hypertension in the right arm with normal to low blood pressure in the lower extremities is classic. Poor peripheral pulses and a weak femoral artery pulse may be found in severe cases. This is associated with Turner's Syndrome (D).

Down's Syndrome (A) is associated with ASD, VSD, AV septal defects.
Adult Polycystic Kidney Disease (B) is associated with berry aneurysms in the brain.
Kleinfelter's Syndrome (C) is usually not associated with cardiovascular abnormalities; infertility is a common presentation.
Marfan's Syndrome (E) is associated with aortic insufficiency.
A 26-year-old female presents to the physician after falling and injuring her hand in a squash game. An x-ray of the patient's hand is shown. The physician notes that the patient is of short stature and obese. In addition, the physician is able to elicit twitching of the facial muscles after tapping the patient's cheek right in front of her ear. The physician suspects an underlying genetic condition.

Which of the following laboratory abnormalities is this patient likely to have as a result of the genetic condition?

a. Excess growth hormone
b. Hypercortisolism
c. Hypoaldosteronism
d. Hypocalcemia
e. Hypophosphatemia

**Correct answer:** D

This patient likely has pseudohypoparathyroidism, an autosomal dominant genetic disorder in which the kidneys are unresponsive to parathyroid hormone, leading to hypocalcemia, and hyperphosphatemia. The cluster of obesity, short stature, shortened 4th and 5th metacarpals and hypocalcemia is characteristic of pseudohypoparathyroidism. The physician was able to elicit a Chvostek sign (twitching of facial muscles after tapping the facial nerve just in front of the ear) which is a sign of hypocalcemia. (Choice D)

Excess growth hormone (Choice A) is likely to be seen in acromegaly, a condition in which the pituitary gland secretes excess growth hormone and causes the liver to produce more IGF. Patients usually present with large hands, feet, increased jaw size and frontal bossing (increased forehead prominence).

Hypercortisolism (Choice B) would be seen in Cushing's syndrome, which could be due to a pituitary adenoma secreting excess ACTH or to primary adrenal hyperplasia, both possibly associated with MEN syndromes. Cushing's is associated with osteoporosis, but characteristic physical findings include truncal obesity, moon facies and purple skin striae, none of which are mentioned in the question.

Hypoaldosteronism (choice C) would be seen in Addison's disease, or primary hypoaldosteronism (Conn Syndrome), in which the adrenal glands are unable to secrete aldosterone. There is an association with skin hyperpigmentation due to Melanocyte Stimulating Hormone.

Hypophosphatemia (choice E) is likely to be seen in hyperparathyroidism, in which the excess parathyroid hormone causes phosphate to be spilled into the urine. Hyperparathyroidism is usually due to a parathyroid adenoma, which could be due to genetic conditions such as MEN I (Multiple Endocrine Neoplasia).
A 20 year old man is advised by family to see a physician after his 33 year old uncle is found to have colon cancer. He has heard from his mother that his grandfather died at age 40 from colon cancer. The patient is referred for colonoscopy which reveals hundreds of pancoonic polyps. A prophylactic colectomy is recommended. A mutation in which of the following genes is most likely to be seen in this patient?

- a. TP53
- b. RB1
- c. APC
- d. BRCA1
- e. BCL2

**correct answer:** C

The correct answer is C (APC).

A mutation of the APC gene, inherited in an AD fashion, results in familial adenomatous polyposis (FAP). The APC gene is a tumor suppressor that prevents nuclear transcription by degrading catenin. In FAP, a mutant APC is inherited, and when the second copy of APC is lost/mutates, colonic polyps develop (“two-hit” hypothesis). Polyp development begins between ages 10 and 20 with malignant transformation often between 35 and 40 years of age. All patients with FAP will develop tubular adenomas and cancer; prophylactic colectomy is recommended.

Choice A (TP53) is a tumor suppressor gene that can be mutated in sporadic colon cancers. The clinical picture given is not consistent with sporadic colon cancer.

Choice B (RB1) is a tumor suppressor gene that inhibits G1 to S phase. Mutations of this gene can result in retinoblastoma, osteogenic sarcoma, or breast cancer.

Choice D (BRCA1) is a tumor suppressor gene that regulates DNA repair. Mutation of this gene can result in breast, ovary, or prostate cancer.

Choice E (BCL2) is a family of antiapoptotis genes. They prevent cytochrome c from leaving the mitochondria, in which it normally activates caspases to initiate apoptosis. Mutation causes increased gene activity which prevents apoptosis. Translocation (8:14), (2:8), (8:22) causes overexpression of BCL2 causing B-cell follicular lymphoma.
A 22 year-old male presents to the emergency department with a swollen and painful right knee. He states that this has happened before, and at home he normally takes factor VIII infusions intravenously three times per week, but he ran out of his medication about one week ago. He states that ice and elevation of the affected joint usually relieve his symptoms but only when he also has his normal factor VIII infusions. His father does not have this disease.

On examination he is afebrile, normotensive and crepitus is appreciated in both knees.

Which of the following is most likely regarding the disorder?

a. Oxytocin is an effective treatment option for many patients because it results in increased levels of factor VIII released from endothelial cells
b. The patient’s sister has a 50% chance of being a carrier for the disease
c. The disease may be caused by parvovirus B19
d. The patient will not pass the mutated allele on to any of his daughter progeny
e. The patient’s father is a carrier for the disease

Correct answer: B

The answer is B. The patient has hemophilia A, which is an X-linked recessive inherited disorder. The mutated allele is on the X chromosome; females are mostly unaffected by this disease and rather may be carriers of the disease. Males are affected when they inherit the abnormal X chromosome from their mother. Thus, female offspring of a female carrier have a 50% chance of being carriers themselves of this disease.

Choice A is incorrect; desmopressin (DDAVP) is a synthetic analog of vasopressin (ADH) which may be used in the treatment of hemophilia A. It acts by increasing the release of factor VIII (and von Willebrand's factor) from endothelial cells. Both oxytocin and ADH are released from the posterior pituitary.

Choice C is incorrect; parvovirus B19 is a single-stranded DNA virus that may cause aplastic crisis in sickle cell patients or fifth's disease ("slapped cheek" syndrome) in children.

Choice D is incorrect; this patient will pass on the mutated X chromosome to all of his daughters

Choice E is incorrect; males are not carriers of X-linked diseases, they are either affected by the disease or they do not have the diseased allele.
A 55 year-old male arrives at your office because recently he attempted to fly for the first time in his life. At the airport, he set the metal detector off even though he was carrying no metal and had no metal implants anywhere. He has no remarkable past medical history or family history that he knows of, although he was adopted. His physical examination is within normal limits. You find that his serum ferritin is >1000 ng/mL. Which of the following is true regarding this disease?

- a. It is caused by an inability to secrete excess copper
- b. Usually, only one copy of the mutated gene is needed to express the phenotype, and the gene is on the X chromosome
- c. Usually, only one copy of the mutated gene is needed to express the phenotype, and the gene is on an autosome
- d. Usually, two copies of the mutated gene are necessary to express the phenotype
- e. The disease inheritance pattern is polygenic

**correct answer:** D

The correct answer is D. The patient has hemochromatosis, which is an autosomal recessive disease. Autosomal diseases require two copies of the mutated allele to be expressed. Hemochromatosis results in increased iron absorption in the small intestine, which can deposit in many places in the body including the liver. Usually women with the disease present later in life because their normal menstrual cycles allow the body to rid itself of some excess iron. Typically, high serum iron levels and high serum ferritin levels occur in this disease. The above case is not an uncommon first presentation for the disease.

Choice A is incorrect; copper metabolism is not affected in hemochromatosis, and ferritin levels reflect iron storage, not that of copper.

Choice B is incorrect; this describes a sex-linked inheritance pattern

Choice C is incorrect; this describes an autosomal dominant inheritance pattern

Choice E is incorrect; hemochromatosis is most commonly caused by mutations in one single gene, HFE3 located on chromosome 6.
A 4-year old child presents to your clinic for a respiratory tract infection. Past history is positive of recurrent pyogenic infections. On examination, he has purpuric and eczematous lesions. A full blood count was done showing small platelets. Which of the following describes the disease best?

a. It is autosomal recessive
b. It is caused by a failure to mount an IgM response against bacterial capsular polysaccharides
c. Immunoglobulins of all classes are low
d. It is a defect in complement
e. It is a defect in microtubule function

**Correct answer:** B

The presentation is typical of Wiskott-Aldrich syndrome, an X-linked recessive disorder characterized by failure to mount an IgM response against bacterial capsular polysaccharides. The mnemonic to remember is WIPE.

1. Wiskott-Aldrich syndrome presents with:
2. Infections (recurrent pyogenic infections)
3. Purpura (thrombocytopenic)
4. Eczema

Option C represents Bruton's agammaglobulinemia (also X-linked recessive)

Option D usually presents according to which component of complement is defective. C1 esterase deficiency - hereditary angioedema. C3 deficiency - recurrent infections (but no eczema therefore this can't be the answer). C6-8 deficiency - Neisseria infections. DAF deficiency - paroxysmal nocturnal hemoglobinuria.

Option E is characteristic of Chediak-Higashi syndrome.
Screening of an African American population in Minnesota yields allele frequencies of $\frac{7}{8}$ for the A globin allele and $\frac{1}{8}$ for the sickle globin allele. A companion survey of 6400 of these people's ancestors in central Africa reveals 4600 individuals with genotype AA, 1600 with genotype AS (sickle trait), and 200 with genotype SS (sickle cell disease—603903). Compared to their descendants in Minnesota, the African population has which of the following?

- A lower frequency of AS genotypes consistent with inbreeding frequency
- A lower frequency of AS genotypes consistent with malarial exposure lower
- A higher frequency of AS genotypes consistent with heterozygote advantage
- A higher frequency of AS genotypes consistent with selection against the S allele
- Identical A and S allele frequencies as predicted by the Hardy-Weinberg law

**Correct answer:** C

Under certain conditions, the Hardy-Weinberg law allows one to interconvert genotype and allele frequencies in a population by using the formula $(p + q)^2 = p^2 + 2pq + q^2$. For a locus with two alleles, p represents the frequency of the more common allele, q of the less common allele, and $p + q = 1$. The Minnesota population therefore has $p^2 = \frac{7}{8} \times \frac{7}{8} = \frac{49}{64}$ (4900 individuals) with the AA genotype, $2pq = 2 \times \frac{7}{8} \times \frac{1}{8} = \frac{14}{64}$ (1400 individuals) with sickle trait (AS genotype), and $q^2 = \frac{1}{8} \times \frac{1}{8} = \frac{1}{64}$ (100 individuals) with sickle cell disease (SS genotype). The African population has a higher frequency of AS and SS genotypes caused by heterozygote advantage for the AS genotype that confers resistance to malaria.
A 6 year old girl is being evaluated for short stature. She is at the 12th percentile for height and the 34th percentile for weight. Vital signs are within normal limits. And physical exam shows widely spaced nipples and a high arched palate. Karyotyping shows 45XO. Which is she most at risk of developing?

a. Bipolar disorder  
b. Breast cancer  
c. Mental retardation  
d. Mitral valve prolapse  
e. Osteoporosis

**Correct answer:** E

The patient’s short height, high arched palate, wide spaced nipples and 45 XO karyotype are all consistent with a diagnosis of Turner syndrome. Patient with Turner syndrome are prone to the development of osteoporosis, which increases the risk of bone fracture. This is due to low estrogen levels from gonadal dysgenesis. This may also be due to having only one copy of X chromosome genes that may be involved in bone metabolism. Estrogen replacement therapy is give to nearly all patients to promote normal maturation, but also has the effect of reducing the risk of osteoporosis.
A 36-year-old male is diagnosed with a rare trinucleotide repeat disorder. He retains full mental ability, but is experiencing progressive weakness, loss of coordination, and an unsteady gait. No other signs on physical exam are found. What is the likely diagnosis?

- Fragile X syndrome
- Friedrick's ataxia
- Huntington's disease
- Myotonic dystrophy
- Spinocerebellar ataxia

**Correct answer:** E

Of each of these diseases, myotonic dystrophy and spinocerebellar ataxia are the only two that don't present with some sort of mental deterioration. Myotonic dystrophy has significant additional signs including muscle wasting, cataracts, and heart conduction defects. Therefore, spinocerebellar ataxia is the likely diagnosis.

Huntington Disease is an autosomal dominant disorder. Characterized by progressive degeneration and atrophy of the striatum and frontal cortex. Clinical abnormalities do not present themselves until 30-40 years of age. Sym. athetoid movements, then progressive deterioration leading to hypertonicity, fecal and urine incontinence, anorexia, and finally dementia and death. Huntington's Disease is marked by increased numbers of CAG trinucleotide repeats within the HD, huntingtin, gene on the short arm of chromosome 4.

(Reference BRS Pathology 2006)

It's important to know two things about each of these diseases: 1) how the disease presents and 2) how it is passed (autosomal dominant vs. recessive; sex-linked). You will NOT need to know the actual trinucleotide repeat (CAG, CTG, etc.). You're guaranteed to see a question like this on the boards!
An infant is noted to have epicanthal folds, a broad nasal bridge, a large tongue, small ears, hypertelorism, Brushfield spots, a single palmar crease on each hand, and a harsh holosystolic murmur. What is the most common cause of this syndrome?

a. Deletion in the short arm of chromosome 5  
b. Maternal meiotic nondisjunction  
c. Paternal meiotic nondisjunction  
d. Robertsonian translocation  
e. Triploidy

**correct answer:** B

This infant has Down syndrome. Trisomy 21 is most commonly caused by maternal meiosis I nondysjunction (the incidence increases with advanced maternal age). Patients with Down syndrome have an increased incidence of congenital heart disease. Hypertelorism refers to wide-spaced eyes. Brushfield spots are small spots at the periphery of the iris formed by abnormal connective tissue aggregation. Triploidy usually results in spontaneous abortion. Deletion in the short arm of chromosome 5 leads to Cri-du-chat syndrome.
A mother brings her two year-old son to your office because he has not yet spoken any words. On exam, he is very quiet, has a prominent jaw, large ears and large testicles. His mother states that she has a brother who is mentally retarded. Cytogenetic analysis reveals a genetic defect to be the cause of the patient's mental retardation. What is the mostly likely mechanism of this genetic defect?

a. Maternal non-disjunction
b. Novel point mutation
c. Paternal non-disjunction
d. Trinucleotide repeat
e. Uniparental disomy

**Correct answer:** D

Fragile X syndrome is the most common cause of mental retardation in males. Key diagnostic factors are lack of milestone attainment, especially language, from a young age, large ears, large testes, a prominent jaw, a long face and flat feet. It is X-linked recessive inheritance that is caused by increased number of CGG repeats on the FMR1 gene. Normal CGG repeats are between 6 and 55, while Fragile X FMR1 alleles have over 230 repeats.
A mother brings in her 15-year-old son to see you. His father recently died suddenly due to a heart condition and she would like you to evaluate her son for the same disorder. On exam, the patient is 6 feet tall with long arms and fingers, an inwardly-curving sternum and severe scoliosis. You refer him for an echocardiogram. Assuming that the father was heterozygous for a genetic disease, what is the probability that his sister will also have this condition?

a. 0%
b. 25%
c. 50%
d. 75%
e. 100%

**Correct answer:** C

This boy has Marfan’s syndrome, an autosomal dominant condition resulting from mutations in the fibrillin-1 gene. Assuming the patient’s mother does not have Marfan’s syndrome, his sister has a 50% chance of having Marfan’s syndrome. If the patient’s mother is affected, 75% of their children will have the syndrome.

Manifestations of the syndrome include arm span exceeding height, arachnodactyly (long, thin fingers), pectus excavatum (deformity of the sternum resulting in a “caved-in” or sunken appearance), dilatation and dissection of the ascending aorta, and ectopia lentis (displacement of the lens upwards). Morbidity and mortality from Marfan’s syndrome is typically related to dilatation and eventual dissection of the ascending aorta, the likely cause of death in this patient’s father.

Hint: Disorders involving structural proteins, such as fibrillin or collagen, tend to be dominant since at least half of the patient’s structural proteins will be compromised. In contrast, disorders involving enzymes tend to be recessive because an individual can typically function with the reduced level of enzyme produced from the normal parental allele.
A biracial couple comes to your office because they are interested in having a child and want to know more about the risks of genetic disease. In particular, the couple is concerned about the risk of cystic fibrosis (CF). The male is of Ashkenazi Jewish descent and has a 1 in 30 CF carrier risk. The female is of Asian descent and has a 1 in 80 CF carrier risk. You advise them that the risk of their child having CF would be exceedingly low, but you recommend a genetic screen if the couple is still concerned. If this couple decides to go forward and have a child, what is the risk of their child having CF?

a. 1 in 600  
b. 1 in 1200  
c. 1 in 2400  
d. 1 in 4800  
e. 1 in 9600

correct answer: E

Each person has a 50% chance of passing on the affected allele. Also, given the differences in their ethnicities, the couple has different risks (probabilities) of being CF carriers.

To determine the child's risk, one needs to multiply by the child's risk of having both the defective genes, one from each parent, which is 1/4.

Thus, you have $\frac{1}{30} \times \frac{1}{80} \times \frac{1}{4} = \frac{1}{9600}$ or 1 in 9600.
**Genotype** | **Count**  
--- | ---  
1-1 | 26  
1-2 | 14  
2-2 | 10

**Assuming Hardy-Weinberg equilibrium, in this restriction fragment polymorphism study of population above what is gene frequency of allele 2?**

- a. .52
- b. .2
- c. .34
- d. .66
- e. .24

**Correct answer:** C

The answer is (C) .34.

The first thing to realize is that just because "Hardy-Weinberg" and "RFLP" are two words that cause diarrhea in most medical students you can ignore than this and focus instead on the what the question is asking.

The Allele frequency is just the proportion of chromosomes that have a specific allele, versus the genotype frequency which measures the proportion of each genotype (this is more general and you are only looking at who who has the gene) in a population.

You have to realize that 26+14+10 is 50 and represents the genotype count total. From there you must see that they are asking for the genotype frequency of allele 2 and you have to take into account the last two genotypes (heterozygous 1-2 and 2-2); because there are two copies of the "2" allele in 2-2 and one copy in 1-2 the calculation of \((2 \times 10) + 14\) / 100. Don't forget that 100 represents the total number of autosomes, because we know that each diploid somatic cell has two copies and there are 50 total and thus 50 x 2 =100. Also, note that we could quickly figure out the allele frequency of Allele 1 because that would be (1-.34) or .66.

- (A).52 would be the genotype frequency of 1-1.
- (B).2 would be the genotype frequency of 2-2.
- (D).66 would be the allele frequency of Allele '1'
- (E).28 would be genotype frequency of the heterozygous 1-2.
Down syndrome occurs more frequently in pregnancies of older mothers, and screening is usually able to identify affected fetuses. These pregnancies often are subsequently terminated.

What accounts for the continued widespread birth of affected individuals?

a. The threshold for screening data is set high
b. CVS samples often are contaminated with maternal cells
c. Pregnancies are more common in younger women
d. Ultrasound is unreliable when a fetus is moving rapidly
e. Karyotype results often are delayed

**correct answer:** C

The answer is C. Although the incidence of Down syndrome clearly rises with maternal age, the number of pregnancies is much higher in younger than in older mothers, and some younger mothers are not screened aggressively for the disorder. The screening thresholds are based on large population studies to both maximize detection and minimize unnecessary testing; lowering them would greatly increase the need for studies, with attendant costs and anxiety (choice A). Contamination of CVS with maternal cells is infrequent (choice B). Ultrasound is occasionally uninterpretable (due to fetal position or movement), but an experienced operator is usually successful. The blood tests also provide another source of information (choice D). Delays in obtaining results of karyotyping are not a problem when rapid methods such as FISH are used (choice E)
The wife of a 48-year-old male patient brings him to the emergency room and says that his memory has progressively gotten worse over the last several years. She also says his personality has been changing. The physician notes abnormal writhing movements of the man’s limbs and hyperreactive reflexes. MRI reveals a loss of volume in the neostriatum and cortex. This disease is inherited via an

a. autosomal dominant trait  
b. autosomal recessive trait 
c. x-linked dominant trait 
d. x-linked recessive trait 
e. 

correct answer: A

the correct answer is A. This patient has Huntington’s disease, which has autosomal dominant inheritance. It is characterized by severe degeneration of the caudate nucleus along with degenerative changes in the putamen and cortex. In addition to chorea, these patients frequently suffer from athetoid (writhing) movements, progressive dementia, and behavioral disorders.
Deletion of a 4 Mb portion of chromosome 15 can result in 1 of 2 possible diseases, based upon which parent this deletion is inherited from. When inherited from the mother, the deletion results in Angelmann Syndrome. If the deletion is inherited from the father, the child will have Prader-Willi Syndrome. This genetic effect is known as:

a. Aneuploidy
b. Anticipation
c. Genomic Imprinting
d. Heredibility
e. Sex-Linked Inheritance

**Correct answer:** C

Genomic imprinting is based on the fact that certain areas of the genome are inactivated based upon which parent the area is inherited from. This is an epigenetic process that relies on methylation and histone modification to activate/inactivate certain areas of the genome.
A patient presents with hypogonadism, increased stature, sterility, and gynecomastia (increased breast tissue). Upon questioning the patient reveals that they were diagnosed at an early age with a chromosomal abnormality resulting in this unique phenotype. Knowing that this is likely due to an aneuploidy, you suspect Klinefelter Syndrome. Which of the following is representative of Klinefelter Syndrome?

- a. 47 XXY
- b. 47 XYY
- c. Trisomy 13
- d. Trisomy 18
- e. Trisomy 21

correct answer: A
Klinefelter's Syndrome results from a XXY.
47 XXY shows increased stature, slight IQ reduction
Trisomy 13 is Patau Syndrome
Trisomy 18 is Edward's Syndrome
Trisomy 21 is Down Syndrome.
A female patient presents with reduced stature, webbed neck, shield-shaped chest, renal malformation, diminished IQ and aortic coarctation. The patient informs you that they were diagnosed as a child with a chromosomal aneuploidy. You suspect:

a. Down Syndrome (Trisomy 21)
b. Edward Syndrome (Trisomy 18)
c. Klinefelter Syndrome (47, XXY)
d. Patau Syndrome (Trisomy 13)
e. Turner Syndrome (45, X)

**Correct answer:** E

The vignette is descriptive of Turner Syndrome which results from a deletion of the SHOX transcription factor gene.
Two genes are expressed, one wildtype and one mutant. The sequences of the gene products are provided above. What is the likely mutation?

- a. Deletion of 6th codon
- b. Deletion of nucleotide in 6th codon
- c. Insertion of stop codon
- d. Substitution of 13th codon
- e. Substitution of 6th codon

**Correct answer:** B

The important points to recognize in the resulting sequence are 1) truncation of more than one residue; 2) change in amino acids from 6th codon onwards. Only a frameshift mutation (nucleotide deletion in 6th codon) explains both the truncation and the change in amino acids.

The important part to note is that this question is giving you the sequence of two gene products, each with a different amino acid sequence. Thus each letter represents an amino acid, not a nucleotide.

Stop codons are UAG, UAA, UGA.
A man with a history of hyperextensible joints and aortic insufficiency presents for a genetic screening. He is discovered to have a homozygous defect in the gene that encodes the glycoprotein fibrillin. What are the chances that his son will have the same disease?

a. 0%
b. 20%
c. 50%
d. 80%
e. 100%

**Correct answer:** E

This man has Marfan syndrome with a characteristic defect in the fibrillin gene. Since he is homozygous and since fibrillin is inherited in an autosomal dominant fashion, his son will certainly be affected as well.

Marfan's gene is FBN-1 (fibrillin) gene on chromosome 15. Fibrillin connects to Extracellular matrix and also TGF-beta. Excess TGF-beta is also proposed as part of the pathophysiological mechanism for Marfan's physiological symptoms.

Marfan's diagnosis: long limbs, dislocated lenses, and aortic root dilation -
A 9 year old girl presents with a history of repeated bone fractures. On physical exam it is noted that her sclerae appear blue, she has reduced hearing ability, and she has small, misshapen, yellowish-blue teeth. Her condition is caused by abnormal synthesis of type-1 collagen. Which of the following is the likely mode of inheritance of this condition?

- a. Autosomal Dominant
- b. Autosomal Recessive
- c. Multifactorial
- d. Not inherited, related to exposure to toxins in utero
- e. X-linked Recessive

**Correct answer:** A

A: The answer is A, autosomal dominant. There are two ways to approach this question. One is the “buzzword medicine” method, by which this case of osteogenesis imperfecta (alluded to by the constellation of symptoms and the pathophysiology of deficient Type I collagen) is known to be one of a few autosomal dominant diseases.

The more reasonable approach to this question is to reason out the mode of inheritance from the scenario. Even though no information is given about the girl’s family members, you can still make a very reasonable prediction about how this disorder is inherited. As the patient is a girl, it is highly unlikely that this disease is, choice E, X-linked recessive.

As the disease affects many organ systems, it is unlikely to be due to, choice D, a toxin exposure at specific stage of in utero development. Disorders resulting from in utero toxin exposure cause lesions at the specific anatomical structures that are being developed during the period of time (holoencephalopathy, cleft palates, polydactyly are examples). The pathophysiology of the disease is too specific to likely be multifactorial (Choice C); this is more common in common diseases such as diabetes and Alzheimer’s. As the disease is presenting relatively late and not in a dramatic fashion (as is seen in some in-born errors of metabolism), it is less likely to be autosomal recessive (Choice B).

Finally, because this is a disorder of structural proteins, it is more likely to be a dominant mutation; enzyme disorders tend to have recessive inheritance.
A couple that is trying to conceive a child once gave birth to a baby that died at 7 months of age. This previous baby was born with severely diminished motor tone and tongue fasciculations. Autopsy revealed massive degeneration of the baby's anterior horns, which was attributed to Werdnig-Hoffman Disease.

What is the probability that another child conceived by this couple will have the same disease?

a. 10%

b. 25%

c. 50%

d. 75%

e. 100%

**Correct answer**: B

Werdnig-Hoffman disease is inherited autosomal recessively. Since each parent is carrying one recessive allele (since one baby has already had the disease), all future children have 25% chance of being recessive homozygous for gene (B).

Werdnig-Hoffman disease is fatal at a very young age, and so is not a negative dominant trait (or the parents would have died), so 100% (E) is incorrect.

For straight-forward autosomal inheritance patterns, you could also eliminate (A) 10%, as no combination of dominant/recessive alleles (based on a Punnett-square analysis) would result in this probability.

Autosomal dominant traits are inherited at a 75% rate (D), while the heterozygous genotype is inherited at a 50% rate (C).
A woman whose father was recently diagnosed with Huntington's disease comes to you to discuss the possibility that her offspring will inherit the disease. Assuming that her mother and her partner have no history of Huntington's disease in their families, and that the father was a carrier for the disease, you tell her that the likelihood her offspring will inherit the disease is which of the following?

- a. 0% or 25%
- b. 0% or 50%
- c. 0% or 100%
- d. 25% or 75%
- e. 50% or 100%

**Correct answer:** A

The gene responsible for Huntington's disease is located on the short arm of chromosome 4, specifically 4p16.3. The disease results when a sequence of three DNA bases is repeated multiple times, qualifying it as one of the trinucleotide repeat disorders. Because Huntington's disease is inherited in an autosomal dominant fashion, only one copy of the mutant allele is necessary for the disease to manifest.

If the woman's father has Huntington's disease and her mother does not, then her chance of having inherited the mutant allele, and manifesting the disease, is 50%. If she does have the mutant allele and her partner does not, than her chance of passing it on to her offspring is also 50%. The overall probability that her child will inherit the mutation based on known information is 50%*50%=25%.

If, however, she has not inherited the allele from her father, than her chance of passing it on is 0%. So depending on wether she inherited the allele or not, her chance of passing it to her offspring is either 0% or 25%.
A 40-year-old woman presents to her primary care physician with a lump in the upper outer quadrant of her left breast. She is diagnosed with invasive breast cancer. Her family history indicates that her mother was diagnosed with breast cancer at age 38, her aunt was diagnosed with ovarian cancer, and a maternal uncle also had breast cancer. The inherited mutation results in alterations in which cellular process?

a. Cell cycle checkpoints  
b. DNA repair  
c. Mitogenesis  
d. Nitrogen base synthesis  
e. Telomere maintenance

**Correct answer:** B

The high frequency, low age of onset, and presence of male breast cancer point to an inherited breast cancer syndrome. The addition of ovarian cancer makes it more likely that this is a mutation in BRCA1 or BRCA2, both of which are involved in DNA repair (B). Mutations in Rb will cause alterations in cell cycle checkpoints (A) and are associated with retinoblastoma. Somatic mutations in genes involved in mitogenesis (C) are related to cancer, but there are no germ-line mutations related to breast cancer. Mutations in synthesis of the DNA bases (D), such as in orotic aciduria will result in immunodeficiency, not cancer. Alterations in telomere maintenance (E) are involved in the immortalization of cancers, but again there is no hereditary cancer syndrome associated with altered telomere maintenance.
A 20-year-old woman presents complaining of amenorrhea. She has never experienced menses. On physical exam it is noted that she has an extremely short stature, little pubic hair, and webbing of the skin on her neck. Which of the following other physical findings is most likely to be seen in the patient?

a. Gait abnormalities  
b. Galactorrhea  
c. Hirsutism  
d. Holosystolic murmur  
e. Multiple painful masses in the breasts

correct answer: D

This patient likely has Turner's syndrome. There are relatively few causes of primary amenorrhea (never had menses) that should always be investigated, and Turner's (pure or mosaic) is relatively common. The short stature and webbing of the neck are highly suggestive of Turner's syndrome, which is a genotype of 45X0. Patients with Turner's syndrome have a variety of symptoms; coarctation of the aorta is one of the more serious and common abnormalities, which can be detected as a holosystolic murmur louder on the patient's back (as opposed to the left lateral decubitus holosystolic murmur of mitral regurgitation).

Heart defects are quite common in chromosomal diseases such as trisomy 21 (Down syndrome), trisomy 18 (Edwards syndrome), and trisomy 13 (Patau syndrome).
A 19 year old girl presented to pediatricians office because she had not began menstruating. The patient found this extremely distressing as her mother and older sister both began their menses at ages 12 and all of her friends had also began their periods. On exam, the girl was 5'4", 145lbs with normal breast development and decreased axillary/pubic hair patterns. Normal female external genitalia was noted. Bimanual pelvic exam revealed a short blind ending vagina without palpable uterus or fallopian tubes. Findings were confirmed by ultrasound. Chromosomal analysis revealed a 46XY karyotype. Which of the following is most likely to account for this patient's presentation?

a. Androgen insensitivity
b. Male pseudohermaphroditism
c. Testicular agenesis
d. Mitochondrial DNA mutation
e. Fragile X syndrome

correct answer: A

Question stem describes a patient with a 46XY karyotype with female external genitalia and secondary sex characteristics. Androgen insensitivity ("testicular feminization") is a type of male pseudohermaphroditism (B) caused by a X-linked inherited defect in the androgen receptor. This causes the cells of the affected individual to be unresponsive to dihydrotestosterone (DHT) but leaves them sensitive to estradiol. Unopposed estrogen exposure causes the fetus to develop phenotypically female, accounting for the normal female external genitalia. Due to the presence of the Y chromosome, however, the fetus will develop male internal genitalia, accounting for the blind ending vagina and lack of uterus or fallopian tubes.

Testicular agenesis (C) does not occur in people with androgen insensitivity syndrome and would not account for the patient's female phenotype. However, lack of normal testicle descent is often seen in the syndrome which often prolongs the diagnosis from being made until late adolescence as in this patient. Mitochondrial DNA mutation (D) would not produce any abnormalities are maternally transmitted and do not involve sex chromosomes or sexual characteristics. Fragile X syndrome (E) is a trinucleotide repeat disorder that is characterized by large or protruding ears, flat feet, macroorchidism and elongated facies. It would not be expected to produce the features seen in this patient.
A 29 year old male presents to your office with his wife because they have been unable to conceive despite having unprotected sexual intercourse for the past 13 months. Physical exam of husband reveals a tall man with mild bilateral gynecomastia, small testicles and reduced body hair. Laboratory studies reveal elevated FSH and a testosterone level that is slightly below normal limits and semen analysis reveals marked oligospermia. Which of the following is most likely to be found on karyotype analysis:

a. 47XXY  
b. 69XXY  
c. 47XYY  
d. 46XY  
e. 47XY + 18

**correct answer**: A

This presentation is classic for Kleinfelter’s Syndrome. Kleinfelter’s syndrome is caused by a XXY karyotype that is formed from nondisjunction during meiosis. Despite having normal male external genitalia, affected men have female secondary sex characteristics (including gynecomastia, soft skin and female hair distribution, hypogonadism (with testicular atrophy and decreased testosterone leading to increased LH and FSH) and tall stature. Kleinfelters syndrome usually remains undiagnosed before puberty and is a common cause of male infertility.

69XXY (B)- karyotype seen in partial hydatidiform moles, composed of 2 paternal and 1 maternal haploid set of chromosomes, usually from the fertilization of the ovum by two or more spermatozoa. Partial moles can contain fetal parts but do not put the patient at increased risk for developing choriocarcinoma, in contrast to complete moles which contain no fetal parts but carry an increased risk of choriocarcinoma.

47XYY (C)- XYY syndrome occurs with increased frequency among criminals with violent behavior. Other characteristics include severe acne, tall stature and rarely mild mental retardation. Gonadal function is normal.

46XY (D)- This is the normal male genotype and would not be expected to produce the characteristics of the patient described in this question stem.

47XY + 18- karyotype associated with Edward’s syndrome (trisomy 18) which results from nondisjunction. Characteristics of include mental retardation, micrognathia, low-set ears, rocker-bottom feet, congenital heart disease and flexion deformities of the fingers. Affected individuals die at a very young age.
A 42 year old female presents to your office with a mass located in the lateral upper quadrant of the left breast. Fine Needle Aspiration reveals a malignancy. Which Oncogene is most likely involved in this neoplasia

- a. BRCA1
- b. BRCA2
- c. Erb-B2
- d. C-myc
- e. ras

**Correct answer:** C

Erb-B2 is the oncogene associated with breast cancer.
BRCA1 and BRCA2 are Tumor suppressor genes
C-myc is associated with Burkitt’s Lymphoma
ras is associated with colon cancer
A 32 year old woman comes to the hospital complaining of acute onset chest pain radiating to her back. She was gardening when the pain began and has remained constant over the past four hours. She has a history of mitral valve prolapse and has been to the emergency room for recurrent joint dislocation. Patient is afebrile, with a heart rate of 97 BP 110/56, weight of 127lb and height of 72in. A CT scan shows the image above.

Patient is subsequently found to have a mutation in the fibrillin gene. What is the most likely mode of inheritance of this condition?

a. Autosomal recessive  
b. Sex-linked recessive  
c. Autosomal dominant  
d. Sex-linked recessive  
e. Mitochondrial inheritance pattern

**correct answer: C**  
C IS THE CORRECT ANSWER.

This question requires that you know the presentation of Marfan’s syndrome. It is an autosomal dominant disease characterized by arachnodactyly, joint and lens dislocation, and aortic dissection and mitral valve prolapse. Individuals are usually tall and thin. Disease is usually asymptomatic and the only presentation may be a sudden aortic dissection as in this case shown in the CT scan. Another similar autosomal dominant disease will be osteogenesis imperfecta, however this is mostly characterized by bluish sclera, hearing loss, dental loss and multiple fractures also known as brittle bone disease.

A IS INCORRECT: Marfan’s syndrome is autosomal dominant  
B IS INCORRECT: Marfan’s syndrome is autosomal dominant  
D IS INCORRECT: Marfan’s syndrome is autosomal dominant  
E IS INCORRECT: Marfan’s syndrome is autosomal dominant
An 18 month old boy is brought to your office with a recent fracture of his wrist and multiple old fractures in the ribs and long bones. You are suspicious of possible child abuse, however to rule out a critical underlying medical condition you perform a complete physical examination of the child. You observe minimal bruising, disorganized teeth, and extreme mobility of joints. The parents of the child deny any history of abuse. You conclude that the boy has a genetic defect. Which biochemical pathway is most likely affected by this defect?

a. Synthesis of fibrillin  
b. Synthesis of type I collagen  
c. Synthesis of type II collagen  
d. Synthesis of type IV collagen  
e. Synthesis of elastin  

**Correct answer:** B
This question presents a boy with the most common variant of osteogenesis imperfecta, which is an autosomal dominant genetic defect in the synthesis of type I collagen. Most cases are mistaken for child abuse unless careful examination is carried out to evaluate for other signs associates with this genetic defect such as blue-tinged sclera, joint hyperextensibility and disorganized teeth.

Defects in synthesis of fibrillin (choice A) causes Marfan’s syndrome, another autosomal dominant disease characterized by arachnodactyly, mitral valve prolapse, joint and lens dislocation. Patients with Marfan’s may present with aortic dissection.

Defects in synthesis of type II collagen (choice C) affects cartilage and vitreous humor of the eye.

Type IV collagen (choice D) is found in basement membranes.

Defects in synthesis of elastin (choice E) is not associated with osteogenesis imperfecta.
A 20 year old man presents to the office complaining of multiple "skin spots" that are scattered across his back, as shown in the image above. He states that he has had them for years, but they are now more numerous and noticeable. Physical exam reveals scoliosis of the back. Slit lamp examination of the eye reveals several small papules on the iris. In consideration of this patient’s most likely diagnosis, what is he at greatest risk of developing?

- a. Cardiac rhabdomyoma
- b. Pectus excavatum
- c. Pheochromocytoma
- d. Renal angiomyolipoma
- e. Renal cell carcinoma

**Correct answer:** C

This patient presents with classic symptoms of neurofibromatosis type I (NF1), also known as von Recklinghausen’s disease. The "skin spots" shown on the back are neurofibromas and can be few or many in number depending on the patient. Scoliosis is one of many skeletal phenomena that can be present in NF1 patients. The small papules present on the iris are Lisch nodules; they are hamartomas pathognomonic of NF1 that do not interfere with vision. Additional signs include cafe au lait spots, peripheral nerve masses that can occur along the route of any peripheral nerve, and gliomas anywhere in the CNS. This patient is most at risk to develop a pheochromocytoma with concurrent hypertension. As a final note, NF1 is of autosomal dominant inheritance, with the mutated gene located on chromosome 17. To learn more, ask about the family history.

Cardiac rhabdomyomas (choice A) are congenital heart tumors that occur in over 50% of patients with tuberous sclerosis, a genetic syndrome characterized by cutaneous lesions, seizures, hamartomas, and mental retardation.

Pectus excavatum (choice B) is a congenital rib and sternal deformity characterized by a sunken anterior chest wall. It can predispose to cardiac and respiratory malfunction. The genetic condition with which it is most commonly associated is Marfan syndrome, a disorder of fibrillin synthesis that leads to various skeletal abnormalities including arachnodactyly, and cardiac abnormalities including mitral valve prolapse.

Renal angiomyolipomas (choice D) are benign tumors comprised of vascular elements, smooth muscle, and adipose tissue. They may occur as one of the manifestations of tuberous sclerosis (see above).

Renal cell carcinoma (RCC, choice E) is the most common type of renal cancer often characterized by the triad of flank pain, abdominal mass, and hematuria. 50% of patients with von Hippel-Lindau disease (VHL) develop bilateral RCC. VHL is a multi-system vascular disorder consisting of hemangioblastomas of the CNS in addition to extra-neural tumors and cysts occurring throughout the body.

Image credit: http://www.opt.indiana.edu/V543/Pathology543/graphics/genetic/G-34.jpg
A sixteen-year-old high school basketball player collapses suddenly after scoring the game winning basket. Despite appropriate resuscitation, the patient dies. An autopsy performed the next day is most likely to reveal which of the following on gross inspection of the heart?

a. An accumulation of fluid within the pericardium compressing the four chambers of the heart
b. A large thromboembolism completely occluding the left anterior descending artery
c. A left atrial myxoma obstructing the mitral valve composed of scattered cells located within a mucopolysaccharide stroma
d. A ruptured chordae tendineae secondary to spontaneous rupture
e. A thickened septum due to hypertrophy and a disorganized and thickened collagen matrix

**correct answer: E**

The correct answer E, is a thickened septum due to hypertrophy and a disorganized and thickened collagen matrix. The patient had hypertrophic cardiomyopathy, an autosomal dominant disorder of the cardiac sarcomere. The thickened and hypertrophied septum leads to narrowing of the left ventricular outflow tract. During systole, the outflow tract is further obstructed secondary to the inward movement of the interventricular septum and this is the mechanism of sudden cardiac death in this disease. Seventy-one percent of patients who die suddenly have no or mild symptoms and only fifteen to twenty percent were participating in moderate to vigorous physical activity at the time of death.

Sudden cardiac death can occur from all of the answer choices listed. However, hypertrophic cardiomyopathy is the most likely diagnosis in this case given the scenario of sudden death in a likely healthy, asymptomatic athlete.

Cardiac tamponade is an accumulation of fluid within the pericardium compressing the four chambers of the heart (A). With acute tamponade, death can occur due to decreased venous return, severely constricted cardiac chambers and sudden fall in cardiac output and blood pressure. However, this is unlikely in a healthy, teenage athlete and hypertrophic cardiomyopathy is a better answer.

Patients with a left atrial myxoma are less likely to die from sudden cardiac death (C). These patients often present with cardiovascular symptoms consistent with mitral valve obstruction and left atrial hypertrophy. Other symptoms may include systemic embolization and constitutional symptoms including fever and weight loss.

Patients with sudden cardiac death from a large thromboembolism occluding the left anterior descending artery are more likely to be older and have risk factors for cardiovascular disease (B). This patient is young and healthy with no known risk factors. Therefore, this is a much less likely etiology.

A ruptured chordae tendineae can occur spontaneously and lead to acute mitral regurgitation and sudden death (D). However, this is very rare. Chordae tendineae rupture is usually secondary to trauma, infective endocarditis, and acute rheumatic fever. Thus, this etiology is less likely to be the cause of this patient's sudden cardiac failure.
_____ nerves can be found joining the SA and AV nodes in the heart

a. A. Vagus
b. B. Phrenic
c. C. Thoracic
d. D. Gastric
e. E. Cervical

correct answer: A

vagus supplies SA and AV nodes
An interventional radiologist is performing a procedure in which real-time imaging of the enteric vessels is important. With the catheter, he enters at the femoral artery, traces back up through the aorta, and enters the celiac trunk. If he were to inject contrast at this point while recording a radiograph, which three vessels would he likely see?

- a. Left gastric artery, short gastric artery, hepatic artery
- b. Left gastric artery, splenic artery, cystic artery
- c. Left gastric artery, splenic artery, common hepatic artery
- d. Right gastric artery, splenic artery, hepatic artery
- e. Superior mesenteric artery, splenic artery, hepatic artery

**Correct answer:** C

The branches of the celiac trunk are the left gastric artery, the splenic artery, and the common hepatic artery.
A 52-year-old male presents with severe epigastric pain and episodes of vomiting blood. The pain is usually relieved by consumption of food or Antacids. The patient's history is significant for prior episodes of epigastric pain for which he receives triple therapy. Which of the following vessels is most likely implicated in the patient's current complaints?

- a. Azygos vein
- b. Gastroduodenal artery
- c. Left gastric artery
- d. Left gastroepiploic artery
- e. Portal vein

**Correct answer:** B

This patient has a past medical history, medication history, and presentation consistent with a bleeding peptic ulcer. He is receiving triple therapy, which indicates that it is suspected his ulcer is related to H. pylori infection. The majority of H. pylori-related ulcers occur in the first or second segment of the duodenum, with the remainder more common in the antrum of the stomach.

Thus, the most likely location of the ulcer is in the duodenum. The gastroduodenal artery, a branch of the common hepatic, courses around the first segment of the duodenum, and is thus the most likely bleeding vessel.

Both the left gastric and left gastroepiploic arteries course along the stomach and may explain the bleeding, but are less likely than a duodenal ulcer.

The portal vein or one of its tributaries may cause bleeding or hematemesis in patients with venous congestion (e.g. cirrhosis, alcoholism); we don't suspect that in this patient.

The azygous vein is retroperitoneal and is extremely unlikely to be causing this patient's GI symptoms in the absence of major trauma.
A 16-year-old girl is brought to the emergency department after attempting suicide by cutting her wrist. The deepest part of the wound is between the tendons of the flexor carpi radialis and the flexor digitorum superficialis. This patient is most likely to have a deficit of which of the following?

- a. Adduction and abduction of the fingers
- b. Extension of the index finger
- c. Flexion of the ring and small fingers
- d. Sensation over the base of the small finger
- e. Opposition of the thumb and other fingers

**Correct answer:** E

This question tests basic anatomical knowledge of the nerves and muscles of the hand and their functions. Such questions are rare on the actual USMLE, but do exist.

The structure between the two tendons mentioned is the median nerve. Loss of the median nerve at the wrist results in loss of thenar muscle innervation, resulting in an inability to oppose the thumb.

(A) Adduction and abduction of the fingers are mediated by the palmar and dorsal interosseus muscles of the hand, respectively. These are mostly innervated by the ulnar nerve.

(B) Extension of the index finger requires forearm extensors.

(C) Flexion of fingers 4 and 5 requires forearm flexor muscles. This is a wrist injury.

(D) The ulnar nerve would mediate sensation over the base of the small finger.

(E) Opposition of the thumb and other fingers involves flexion of the flexor carpi radialis (FCR) and flexor digitorum superficialis (FDS), both innervated by the median nerve.

The FDS muscle in the forearm gives rise to 4 tendons that pass through the flexor retinaculum of the carpal tunnel and insert at the proximal interphalangeal joints. (So, it’s an extrinsic muscle of the hand.)

The FCR muscle passes a tendon just lateral to the FC-ulnaris tendon, and inserts at the base of the 2nd metacarpal.
While playing squash, a young woman tore one of her right rotator cuff muscles. Which of the following is most likely to have been injured?

a. Infraspinatus
b. Subscapularis
c. Supraspinatus
d. Teres major
e. Teres minor

correct answer: C
The supraspinatus is the most commonly injured rotator cuff muscle. The infraspinatus, teres minor, and subscapularis are the other rotator cuff muscles; they could also be torn, but are torn less frequently. The teres major is not part of the rotator cuff.

Supraspinatus muscle initiates abduction
Infraspinatus muscle externally rotates arm
teres minor adducts and externally rotates arm
subscapularis muscle internally rotates and adducts arm

Common mnemonic is SITS: from counter clockwise to clockwise around glemoral humerus: Supraspinatus (inferior to coracoid and acromion), infraspinatus, teres minor, subscapularis (anterior and inferior to supraspinatus)
A high school football player suffers a traumatic knee injury after an opposing player dives into the lateral aspect of his knee. Which structure(s) in the knee are most vulnerable in such an injury?

a. Anterior cruciate ligament, medial collateral ligament, medial meniscus
b. Anterior cruciate ligament, medial collateral ligament, posterior cruciate ligament
c. Anterior cruciate ligament, lateral collateral ligament, posterior cruciate ligament
d. Anterior cruciate ligament, medial meniscus, posterior cruciate ligament
e. Lateral collateral ligament, medial collateral ligament, posterior cruciate ligament

correct answer: A

A common injury resulting from lateral-to-medial knee trauma is injury to the "O'Donoghue tear," or "Unhappy Triad," which consists of the medial collateral ligament, the lateral meniscus, and the anterior cruciate ligament.
A 25-year-old man who has suffered blunt trauma to his head has a suspected injury to his superior rectus muscle. What eye movements might be compromised if this indeed were the case?

a. Abduction only
b. Abduction and elevation
c. Adduction and depression
d. Adduction and elevation
e. Extorsion and elevation

**Correct answer:** D

Simple definition question but important! The superior rectus muscle is a simple bridge-like muscle above the eye that operates dorsomedially and so results in elevation and adduction of the eye. The superior oblique muscle is responsible for depressing and abducting the eye while the inferior oblique muscle is responsible for elevating and abducting the eye. See this link for description of muscle actions: [http://en.wikipedia.org/wiki/Extraocular_muscles#List_of_muscles](http://en.wikipedia.org/wiki/Extraocular_muscles#List_of_muscles)
A 27-year-old man is brought to the emergency room after sustaining a knife wound in the back. His vital signs are within normal limits. On exam, you appreciate a 4 cm long laceration that runs parallel to the inferior medial border of the scapula, and is 2 cm from the spinous process of T5. The laceration has visibly penetrated at least 1 cm into the muscle tissue. Which muscle is most likely affected?

- a. Deltoid
- b. Infraspinatus
- c. Levator scapulae
- d. Rhomboid major
- e. Triceps brachii

**Correct answer: D**

The insertion of the rhomboid major (D) is the medial border of the scapula (it arises from the spinous processes of T2 to T5, as well as the supraspinous ligament). The deltoid (A) originates at the spine of the scapula and goes laterally to insert on the humerus. The infraspinatus (B) originates at the posterior scapula and goes laterally to form part of the rotator cuff (remember: SITS). The levator scapulae (C) inserts at the superior medial border of the scapula and goes superiorly. The triceps brachii (E) originates at the lateral scapula and inserts on the humerus.
A 30-year-old man sustains significant shoulder trauma during a motor vehicle accident. Other than ligamentous damage, x-ray shows only an avulsion fracture of the tip of the coracoid process. Which shoulder movement would be expected to be most impaired?

a. Abduction
b. Extension
c. External rotation
d. Flexion
e. Inversion

Correct answer: D

The coracobrachialis and the short head of the biceps brachii originate at the coracoid process. An avulsion fracture (where a small piece of bone is broken off while still attached to tendon or ligament - in this case tendon) of the coracoid process would prevent these muscles from exerting their main action at the shoulder joint, which is flexion (D). Other muscles responsible for shoulder flexion include the anterior deltoid and the pectoralis major. Supraspinatus and deltoid are responsible for shoulder abduction (A). Shoulder extension (B) is mostly due to the latissimus dorsi and the posterior deltoid. External rotation (C) is mostly due to teres minor, infraspinatus, and posterior deltoid. Inversion (E) is not an action that occurs at the shoulder (it occurs in the ankle).

Memorizing all of these actions is unnecessary - knowing the location of the coracoid process (palpable on exam near the deltopectoral triangle) is sufficient in this case. The question stem is asking for the effect on the shoulder, so (A), (B), and (C) would not be possible given the anterior location of the coracoid process in relation to the humerus.

Shoulder motion can be confusing:

Shoulder flexion is the motion where the arm is raised up (toward superior direction, as in lifting a child).

Adduction is always a motion towards the body, shoulder adduction occurs when you slapping someone across the face.

Abduction is moving away from the body. shoulder abduction is opposite of above - you went to backslap that person with the same hand...
A 22-year-old intoxicated male presents with a painful headache following a bar fight. Head CT reveals a biconvex mass adjacent to a fractured right temporal bone. The artery implicated in the injury passes through the cranium through the same foramen as which of the following nerves?

a. Maxillary nerve
b. Meningeal branch of the mandibular nerve
c. Meningeal branch of the maxillary nerve
d. Middle meningeal nerve
e. Nervus intermedius

**Correct answer:** B

A biconvex (lens-shaped) mass is strongly indicative of an epidural hematoma, which most commonly results from injury to the middle meningeal artery secondary to temporal bone fracture. The middle meningeal artery enters the cranium through the foramen spinosum together with the meningeal branch of the mandibular nerve (B), which is also called nervus spinosum.

The maxillary nerve (A), the V2 division of the trigeminal nerve, courses through the foramen rotundum, not the foramen spinosum.

The meningeal branch of the maxillary nerve (C), also called the middle meningeal nerve (D), does not pass through a foramen in the skull. It branches directly off of V2 immediately after its origin from the trigeminal ganglion.

The nervus intermedius (E) enters the internal auditory canal, not the foramen spinosum. Nervus intermedius should not be confused with nervus spinosum, which is another name for the meningeal branch of the mandibular nerve.
A 23 year old man is brought to the Emergency Department after being involved in a motor vehicle accident. The patient reports not being able to move or feel his legs. You suspect a spinal cord injury. Which of the following best describes the type of joint found between the bodies of the 4th and 5th thoracic vertebrae?

a. Diarthrosis
b. Suture
c. Symphysis
d. Synchondrosis
e. Syndesmosis

Correct answer: C

In general, joints can be classified into two main groups: diarthrosis and synarthrosis.

Synarthrosis are joints that offer little to no movement under normal conditions.

Synarthrosis can be sub-classified as sutures, synchondrosis, or syndesmosis. Syndesmosis (choice E) is a joint where two bones are attached by ligaments but without synovia or articular cartilage.

A symphysis (choice C) is a type of syndesmosis in which a fibrocartilaginous disc is present between the two bones. The bodies of vertebrae are connected by 5 different spinal ligaments as well as a fibrocartilaginous disc.

Sutures (choice B) describe joints of only fibrous connections as seen between certain cranial bones.

Synchondrosis (choice D) describes joints where the connecting material is cartilage, such as the joint between ribs and the sternum.

Diarthrosis (choice A) are true synovial joints whose components always include a connective tissue capsule, a synovial membrane, and hyaline articular cartilage. The joints found between the articular processes of vertebrae are one example.
Which of the following structures is most likely to be injured by a fracture of the posterior cranial fossa?

- a. Abducens nerve in cavernous sinus
- b. Facial nerve at internal acoustic meatus
- c. Maxillary nerve
- d. Occipital lobe of cerebral hemisphere
- e. Temporomandibular joint

**Correct answer:** B

Choice B (facial nerve at internal acoustic meatus) is correct. The internal acoustic meatus is a feature of the posterior cranial fossa. It transmits the facial nerve (CN VII) and the vestibulocochlear nerve (CN VIII).

Choice A (abducens nerve in cavernous sinus) is incorrect because it is in the middle cranial fossa.

Choice C (maxillary nerve) exits the foramen rotundum in the middle cranial fossa.

Choice D (Occipital lobe) is unlikely to be injured.

Choice E (Temporomandibular joint) is a joint between the temporal bone and mandibular and not part of the posterior cranial fossa.
A 30 year old female with no medical history reports episodes of numbness and tingling in both hands for the past 4 months. She has trouble using her keyboard at work. Her first three digits on each hand bother her the most and she recalls no history of trauma. The fingers aren’t painful or swollen. Physical exam shows decreased sensation to light touch and pinprick on the palmar surface of the first three fingers of each hand. Thenar atrophy is noted. Which of the following is the likely cause of her problem?

a. Repetitive stress injury  
b. Diabetes mellitus  
c. Amyotrophic lateral sclerosis  
d. Zoster virus infection  
e. Brachial plexus injury  

**Correct answer:** A  
Choice A is the correct answer  
This patient has carpal tunnel syndrome which is a form of compression neuropathy. It results from entrapment of the median nerve beneath the flexor reticulum. The most common cause is repetitive use and is more common in women. The problem is often bilateral.  
Choice B leads to a symmetric distal sensory neuropathy. It is unlikely given her lack of symptoms of diabetes and distribution of her injury  
Choice C would result in progressive symmetric weakness from loss of motor neurons  
Choice D results in a painful neuropathy in a dermatomal distribution.  
Choice E is unlikely due to lack or trauma and bilateral distribution
A 68-year-old man arrives at your family practice clinic complaining of a week of left-sided ear pain. For the last two days, he has been drooling from the corner of his mouth. Physical exam reveals a left-sided facial weakness. Otoscopic examination is significant for a small cluster of vesicles in the left external auditory canal. What other associated symptom would you expect the patient to report?

- a. Altered sense of smell
- b. Altered taste of food
- c. Deviation of tongue to the left
- d. Difficulty hearing with the right ear
- e. Loss of vision in the left eye

**Correct answer:** B

Ramsay-Hunt Syndrome, or Herpes Zoster Oticus, is caused by the reactivation of VZV in the geniculate ganglion. It classically presents with an ipsilateral Bell’s palsy, ear pain, and clusters of clear or hemorrhagic vesicles in the external ear canal. It is frequently associated with a loss of taste in the anterior portion of the tongue.

While you may not know specifically about Ramsay-Hunt Syndrome, you should be able to reason that the patient has a problem with their CN-VII. Recall that the geniculate nucleus (responsible for controlling CN-VII) is also the termination for the frequently-tested chorda tympani nerve, responsible for conveying taste from the anterior two-thirds of the tongue.

Because zoster reactivates in only one dermatome (here, a cranial nerve), we would not expect to see problems with CNs I (smell), II (vision), XII (tongue deviation), or the contralateral CN-VIII (hearing in the RIGHT ear).
A patient presents to your clinic with a complaint of visual difficulties. On physical exam, you notice that when the patient looks to the left, only his right eye moves. The rest of the exam was within normal limits. The nerve responsible for this patient's problem travels through which of the following structures?

- a. Internal auditory meatus
- b. Foramen ovale
- c. Foramen rotundum
- d. Optic canal
- e. Superior orbital fissure

**Correct answer:** E

The patient in the question stem presents with lateral rectus palsy on the left side. The nerve responsible for this muscle is the abducens nerve (cranial nerve VI). Due to its long path, this cranial nerve is the most susceptible to injury. During its path from the brainstem to the extraocular muscles, the abducens passes through the superior orbital fissure. Other nerves that pass here include cranial nerves III, IV, and V1. Cranial nerve V2 passes through the foramen rotundum. Cranial nerve V3 passes through foramen ovale. The optic nerve, ophthalmic artery and central retinal vein pass through the optic canal. Cranial nerves VII and VIII pass through the internal auditory meatus.
A 2 day old newborn is found to have persistent adduction and internal rotation of his left arm with extension at the elbow, pronation of the forearm, and flexion at the wrist. The delivery of the baby was complicated by a vaginal delivery in which the baby's shoulder was caught on the mother's sacrum. The obstetrician was required to pull hard on the baby's head during delivery. Which nerve roots of the brachial plexus are most likely to be damaged in this situation?

a. C3, C4  
b. C4, C5  
c. C5, C6  
d. C7, C8  
e. C8, T1

**Correct answer:** C

The brachial plexus is composed of nerve roots C5-T1 which form trunks composed of roots C5-C6, C7 and C8-T1. In this situation, the baby has a upper brachial plexus injury. It is caused when there is significant traction put on the upper trunk of the brachial plexus. The nerve roots affected are most commonly C5 and C6, and much less frequently, C7. There is an increased risk of developing Erb-Duchenne palsy when a baby is delivered in the breech position, yanking on the shoulder (either in during delivery or in a situation when a falling person grabs a branch/bar to stop their fall) or when there is a traumatic fall on the side of the neck. The description of the baby's arm in the question stem results from a paralysis of the abductors of the arm, lateral rotators of the shoulder, and the biceps.

Another kind of traumatic palsy is Klumpke’s palsy (thoracic outlet syndrome) is caused by an embryologic defect compressing the inferior trunk of the brachial plexus. The effected nerve roots are C8 and T1 (choice E) and can present with atrophy of the thenar and hypothenar eminences, sensory deficits on the medial side of the forearm and hand, and disappearance of the radial pulse upon moving the head toward the opposite side.

Nerve roots C3-C5 (choices A and B) are involved in the innervation of the of the diaphragm ("3,4,5 keeps the diaphragm alive"), but C3 and C4 are not nerves innervating muscles of the arm.
A 45 year-old male comes to his primary care physician because he is having difficulty moving his head and neck. On examination, the physician notes the patient has decreased strength and motion in the muscles of the neck. His balance is unaffected, he has a normal gait, and the tone in the rest of his limbs is within normal limits. Which of the following is the most likely pathway affected that might cause this deficit?

- a. Lateral vestibulospinal tract
- b. Medial vestibulospinal tract
- c. Medial reticulospinal tract
- d. Lateral reticulospinal tract
- e. Rubrospinal tract

**Correct answer:** B

The answer is b. The medial vestibulospinal tract arises from the medial vestibular nucleus and descends in the MLF to cervical levels, where it controls lower motor neurons (LMNs), which innervate (flexor) muscles controlling the position of the head.

The lateral vestibulospinal tract (answer A) extends down to the lumbar region and is involved with posture by controlling the extensor muscles of the legs.

The reticulospinal tracts (answers C and D) modulate muscle tone of the limbs.

The rubrospinal tract (answer E) facilitates flexor motor neurons of the limbs.
A physician is performing a cranial nerve exam on a patient. While testing the gag reflex, it is noted that when the right side of the pharyngeal mucosa is touched, the patient’s uvula deviates to the right. When the left side is touched, the Pt. does not gag. Which of the following is the most likely location of his lesion?

- a. Left glossopharingeal n. and left vagus n.
- b. Left glossopharingeal n. only
- c. Left vagus n. only
- d. Right glossopharingeal n. and right vagus n.
- e. Right glossopharingeal n. only

**Correct answer:** A

The gag reflex requires the glossopharingeal n. for the sensory limb of the reflex (unilateral) and the vagus n. for the motor limb of the reflex (bilateral). A lesion of the left glossopharingeal n. will denervate the sensory receptors on the left side of the pharynx. Thus when the left side is touched, the Pt. does not feel it and does not gag. The gag reflex requires the vagus n. for motor limb of the reflex. If the vagus n. is lesioned, the left side of the soft palate will not elevate during a gag and the uvula will deviate to the right. In this case, the Pt. only feels the touch on the right side and only elevates the right side of the palate. Thus there is a lesion of both the left glossopharingeal n. and left vagus n.

B. If the Pt. had a lesion of the left glossopharingeal n. only, there would have been no gag when the left side is touched, but there would be a normal gag, without deviation of the uvula, when the either side was touched.

C. If the Pt. had a lesion of the left X n. only, the Pt. would have deviation of the uvula to the right when a gag was elicited, but touching either side of the pharynx would elicit a gag.

D. If the Pt. had a lesion of the right glossopharingeal n. and right vagus n., touching the right side of the pharynx would not elicit a gag and touching the left side would elicit a gag with the uvula deviating to the left.

E. If the Pt. had a lesion of the right glossopharingeal n. only, there would be no gag when the right side is touched and normal gag when the left side is touched.
Lesions involving Brodmann's Area 5 & 7 of Dominant Hemisphere are associated with everything except:

a. Apraxia
b. Loss of Tactile and Proprioceptive sensation
c. Astereognosia
d. Impaired Integration of visual and somatosensory information
e. Aphasia

correct answer: B
An angiographic study of the distal part of the circle of Willis requires access to the vertebral artery. Access to this artery can be obtained via the

- a. anterior triangle of the neck
- b. muscular triangle of the neck
- c. posterior triangle of the neck
- d. submental triangle
- e. suboccipital triangle

**Correct answer:** E

The correct answer is E. The suboccipital triangle lies in the neck at the base of the skull. The suboccipital triangle is bounded by the inferior oblique, rectus major, and superior oblique muscles. The vertebral artery can be found within the suboccipital triangle, lying on the posterior arch of the atlas, lateral to the midline, typically at about the level of the lower portion of the ear lobe. Care should be taken in accessing the vessel, since the first cervical nerve lies between the vertebral artery and the posterior arch of the atlas.

The anterior triangle of the neck (choice A) is in the anterior neck, and is bounded by the neck midline, the mandible, and the sternocleidomastoid muscle. The anterior triangle of the neck is subdivided into the muscular, carotid, submandibular, and submental triangles.

The small muscular triangle (choice B) lies between the superior belly of the omohyoid and the sternohyoid muscle.

The posterior triangle of the neck (choice C) is on the side of the neck, and is bounded by the sternocleidomastoid muscle, trapezius, and the middle third of the clavicle.

The submental triangle (choice D), also called the suprahyoid triangle, is found below the lower jaw, and is bounded by the hyoid bone and anterior bellies of the digastric muscles.
Several members of a large family are affected by the onset of decreasing mental function and motor coordination when they reach middle age. Their movements are marked by choreoathetosis. Genetic testing reveals increased trinucleotide CAG repeats. Which of the following structures is most likely to appear grossly abnormal at autopsy of the affected persons?

a. Caudate nucleus  
b. Midbrain  
c. Temporal lobe  
d. Locus cerelus  
e. Dorsal root ganglion

**Correct answer:** A  
(A) CORRECT. Huntington disease (HD) is inherited in an autosomal dominant pattern. The gene is on chromosome 4, coding for a protein called huntingtin. Normally, there are 11 to 34 copies of the CAG repeat. There are more copies with HD; a greater number of copies correlates with earlier onset of the disease.
A 56-year-old woman complains of colicky right flank pain of several hours duration. She has a history of renal calculi, and denies dysuria. Urinalysis shows microscopic hematuria but no leukocyte esterase. Abdominal x-ray demonstrates a calculus at vertebral level L2.

Where is the renal calculus most likely located?

a. Corticomedullary junction  
b. In the ureter as it crosses the pelvic brim  
c. Intersection of renal pelvis and ureter  
d. Junction of the ureter and bladder  
e. Minor calyx of the kidney

**Correct answer:** C

This patient has a recurrence of kidney stones. Although calculi are often lodged at the junction of the ureter and bladder, in the ureter as it crosses the pelvic brim, or the intersection of the renal pelvis and ureter, the last option correlates with the radiographic finding.

The corticomedullary junction is the site of ascending and descending loops of Henle, as well as collecting ducts. Calculi are unlikely to impact here.

The minor calices of the kidney open into the major calices, and are an unlikely site of impaction.
A 24 year old hispanic man is involved in a motorcycle accident and sustains severe trauma to his external genitals. The emergency department physician is concerned that the urethra may be damaged in the bulb of the penis. If this were the case urine will extravasate into which of the following structure.

a. Anterior thigh  
b. Ischioanal fossa  
c. Peritoneal cavity  
d. Pudental canal  
e. Superficial perineal space

**correct answer: E**

Damage to the urethra in the bulb of penis could result in the extravasation of urine into the superficial perineal space. From there it could gain access to the scrotal sacs and the potential space between the superficial and deep fascias of the penis.

Urine could extravasate into Anterior thigh (choice A) if urethral rupture occurred in the superficial peritoneal pouch.

Urine could extravasate into peritoneal cavity (choice C) if the ureter were damaged.

Urine would not enter Ischioanal (choice B) or Pudental canal (choice D) during urethral rupture.
A 25 year old man comes to the emergency room complaining of acute onset of fever and testicular pain. He also noted that his scrotum had begun to swell and had a moderate amount of erythema. On physical exam, he is found to be febrile to 40 degrees Celsius and has significant bilateral swelling of the scrotum and bilateral testicular tenderness on palpation. Lab values were significant for leukopenia with relative lymphocytosis and a slightly elevated amylase. A high degree of suspicion for mumps prompted antibody testing. The patient was found to be IgM positive for the mumps antibody. What is the primary lymphatic drainage of the testes?

- a. Common iliac nodes
- b. External iliac nodes
- c. Inguinal nodes
- d. Internal iliac nodes
- e. Para-aortic nodes

**Correct Answer:** E

The primary lymphatic drainage for the testes and ovaries are the para-aortic lymph nodes (choice E) located in the lumbar region of the abdominal aorta. The para-aortic lymph nodes are sometimes referred to as the retroperitoneal lymph nodes.

External iliac lymph nodes (choice B) drain external genitalia, vagina, and cervix. Internal iliac lymph nodes (choice D) drain the cervix, prostate, and rectum. The common iliac lymph nodes (choice A) receive afferents from both external and internal iliac groups.

Inguinal lymph nodes (choice C) drains the lower part of the anal canal as well as part of the external genitalia.
An infant born at 27 weeks develops respiratory distress syndrome. In the lungs, alveoli are maintained in their open, expanded state with the help of pulmonary surfactant which helps to reduce surface tension. Which of the following describes the composition of pulmonary surfactant after the 36th week of pregnancy in normal development?

- a. Surfactant with a high proportion of sphingomyelin
- b. A lecithin-to-sphingomyelin ratio less than 2:1
- c. Surfactant containing less-surface-active alpha-palmitate, beta-myristate species
- d. Surfactant containing phosphatidylglycerol
- e. 75% lecithin and 10% sphingomyelin

correct answer: D

Pulmonary surfactant after the 36th week of pregnancy contains phosphatidylglycerol and this is the best sign of maturity of the fetal lungs. In addition, a lecithin-to-sphingomyelin ratio of greater than 2:1 is a marker that the fetus will not develop respiratory distress syndrome and is seen later in the third trimester. Mature pulmonary surfactant contains 75% lecithin and 10% phosphatidylglycerol, not sphingomyelin.

Immature pulmonary surfactant contains a higher proportion of sphingomyelin and the species of lecithin that is less surface active, alpha-palmitate, beta-myristate.
A 51-year-old man comes to the doctor’s office with shortness of breath and productive cough. The patient has a history of smoking for the past 32 years and in the past was told that he had chronic bronchitis. Which of the following structures is most likely to be altered in this patient’s lungs?

a. Alveoli  
b. Bronchial arteries  
c. Bronchial submucosa  
d. Terminal bronchioles  
e. Trachea

_correct answer: C_

The bronchial submucosa (choice C) is enlarged in patients with chronic bronchitis secondary to hyperplasia and hypertrophy of the mucous cells. Also, in the bronchial submucosas there is a greater proportion of mucous cells to serous cells.

The terminal bronchioles (choice D) are destroyed in emphysema leading to exertional dyspnea and nonproductive cough.

The alveoli (choice A) are not altered greatly in chronic bronchitis, but you will see altered alveoli in acute respiratory distress syndrome and acute eosinophilic pneumonia.

The trachea (choice E) and the bronchial arteries (choice B) are not altered in chronic bronchitis.
A 21 year old man goes to his primary care doctor after experiencing dry, scaly skin for several months. He has patches red, thickened skin with areas of silver scaling over his elbows, knees, and scalp. He has had some itching at those areas but over the counter lotion use has not helped much. Histological examination of one of the lesions would most likely show increased thickness in which of the following layers in the photo?

- a. Stratum Corneum
- b. Stratum Lucidum
- c. Stratum Granulosum
- d. Stratum Spinosum
- e. Stratum Basale

**Correct answer:** D

The correct answer is D.

D is the stratum spinosum. It is often increased in thickness in psoriasis, as this individual most likely has based on clinical description.

A is the stratum corneum. Histological examination of this layer in psoriasis will show parakeratotic scaling in which nuclei are still present. Normal stratum corneum should not have nuclei. This layer also will show hyperplasia (acanthosis). Neutrophil collections called Munro microabscesses can be found in this layer with psoriasis.

B is the stratum lucidum.

C is the stratum granulosum. It is decreased in size in psoriasis.

E is the stratum basale. In psoriasis histology shows extension of rete pegs, which are downward extensions of the basale layer into the dermis.
75 year old male professional golfer presents to his physician with a lesion on the lateral aspect of his nose that has been present for several months. When questioned about sun exposure he states during his playing career he rarely wore a hat or sunscreen. On exam, the lesion is papular with rolled edges and appears pearly. This lesion most likely arose from which of the following?

- stratum corneum
- melanocytes
- stratum granulosum
- stratum spinosum
- stratum basale

**Correct answer:** E

The correct answer is E (stratum basale).

The patient most likely has basal cell carcinoma as a result of many years of unprotected sun exposure. BCC is the most common skin cancer and arises from the basal layer of cells in the epidermis. It is common in sun exposed regions such as the head and neck. It is typically described as a pearly, smooth papule with rolled edges and surface telangiectasias. Metastasis is rare but it can be locally destructive.

Choice A (stratum corneum) consists of keratinized, dead cells and contain no nuclei. It is not the source of basal cell carcinoma. Choice B (melanocytes) are the source of melanoma. This is an aggressive skin cancer and number one cause of skin cancer death. Features of melanoma include asymmetry, border irregularity, color variation-pink to black to blue, >5mm in diameter, elevated (ABCDE). Choice C (stratum granulosum) are where keratin proteins and water-proofing lipids are produced and organized. It is not the source of basal cell carcinoma. Choice D (stratum spinosum) is where keratinization begins. This layer is not the source of basal cell carcinoma but can increase in size in psoriasis. The second most common cause of skin cancer is squamous cell carcinoma. It arises from epidermal cells undergoing keratinization but is not associated with any particular layer. Sunlight is the most important risk. Typically it is a crusting, ulcerated nodule or erosion.
Cardiac muscle differs from skeletal muscle in that

a. Cardiac muscle contains myosin light chain kinase and no actin filaments
b. Cardiac muscle lacks the intercalated discs present in skeletal muscle
c. Cardiac muscle forms dyads between the T tubules and skeletal muscle while skeletal muscle forms triads
d. Cardiac muscle requires only sodium for contraction to occur
e. Cardiac muscle is striated while skeletal muscle is not

**Correct answer:** C

Look at a picture of the structure of T tubules and the sarcoplasmic reticulum in cardiac vs. skeletal muscle cells to better visualize the relationship.
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In cardiac muscle the t-tubule form a dyadic contact with the reticulum at the z line while in the skeletal muscle the t-tubule form a triadic contact with the reticulum at the A-I junction.
A 45-year-old man cannot leave home without antacids because he has had severe heartburn with every meal for 10 years. Which of the following is likely to be seen on biopsy of his lower esophagus?

a. Columnar metaplasia 
b. Hyperplasia  
c. Hypertrophy 
d. Normal esophageal epithelium  
e. Squamous metaplasia 

**Correct answer:** A

Barrett’s esophagus is a complication of gastro-esophageal reflux disease (GERD). The lower 2/3 of the esophagus is composed of stratified squamous epithelium. With repeated exposure to caustic gastric acid, tissue in the lower esophagus is replaced with columnar glandular epithelium with goblet cells resembling glandular cells of the intestines that secrete mucin. This process is called metaplasia, in this case columnar.

Hyperplasia is an increase in the number of cells in a tissue region, such as endometrial hyperplasia. Hypertrophy describes enlargement of existing cells, such as in hypertrophic cardiomyopathy. Squamous metaplasia would not occur in the esophagus, where epithelium is normally squamous. Squamous metaplasia can occur in the bladder, cervix, or lungs.
A 44 year old man has diarrhea, foul-smelling stools, cramping, fever, and joint pains. Six months ago his weight was 165 pounds. On this visit he weighs 145 pounds. A slide from a biopsy is shown above.

What is the most likely finding in this patient?

- Histologic findings limited to the duodenum and jejunum
- Antibodies directed against gliadin fraction of gluten
- Herpetiform lesions on the extensor surface of his forearms
- Bacteria-laden macrophages with bacteria that are directly visible only on EM
- Flagellated protozoa with two nuclei

correct answer: D
The correct answer is (D). The picture shows Periodic acid-Schiff (PAS) positive staining of macrophages in the epithelium and lamina propria in a patient infected with Tropheryma whippelii. This organism causes Whipple’s disease, which manifests with malabsorption, arthralgias, and cardiac and neurologic symptoms. PAS staining is used to identify glycogen in tissues, and it stains positive in Whipple’s disease because whippelii bacteria have glycoprotein in their cell walls. Answers (A), (B), and (C) are more likely to be seen in a patient with celiac sprue. Histologic specimens of the proximal bowel would demonstrate blunted villi. Answer (E) refers to Giardia, a protozoan which does not infiltrate the small bowel.
A 16 year girl dies in a drunk driving accident. On autopsy, a sample of her tissue is observed under light microscopy.

Which of the following statements is true regarding the parenchymal tissue shown above?

a. Area II is likely to suffer ischemic injury before area I
b. Area II is likely to be injured by alcohol abuse before area I
c. Area I would be more likely to suffer toxic injury
d. Bile flows from area II to area I
e. The parenchymal tissue shown above is separated from blood cells by fenestrated endothelium with a porous basement membrane

correct answer: C

The correct answer is (C). Area I marks the hepatic sinusoids surrounding the central vein, also known as the pericentral zone, whereas area II marks the sinusoids surrounding the portal triad, known as the periporal zone. Answers (A) and (B) are incorrect. The pericentral zone is the least oxygenated area of liver parenchyma. Thus, it is most likely to suffer ischemic injury. The pericentral zone is also most likely to be injured by alcoholic hepatitis, which usually follows bouts of heavy-drinking. The pericentral zone also contains the cytochrome P-450 enzymes, which can produce toxic metabolites and reactive oxygen species. Alcohol induces P-450 enzymes. Answer (D) is incorrect because bile flows toward the canals of Hering in the periportal area and collects in the bile duct of the portal triad. Answer (E) is incorrect. Although the sinusoids are lined by a fenestrated endothelium, they do not lie on a basement membrane.
A 19-year-old female college student presents to the student health center complaining of a sore throat and headache. She reports that she had been feeling tired and worn-down for the past few weeks, but attributed it to her busy schedule at school. She denies feeling sad or depressed and denies weight loss. On physical exam, she has enlarged posterior chain cervical lymph nodes. What histology is likely to be seen when the physician looks at her peripheral blood smear?

a. All stages of granulocyte maturation
b. Atypical lymphocytes with clumped chromatin and irregular nuclei surrounded by normal RBCs
c. Knobs on RBC membranes and intrerythrocyte organisms
d. Microcytic, hypochromic red blood cells
e. Microspherocytes

**Correct answer:** B

This patient has a classic presentation for mononucleosis secondary to infection with Epstein-Barr virus (EBV). The finding on peripheral blood smear is atypical lymphocytes with clumped chromatin and irregular nuclei. (choice B).

Choice A refers to a neoplastic process such as chronic myelogenous leukemia, in which all stages of granulocyte maturation can be in seen in the peripheral blood (as opposed to just the bone marrow). The typical patient would present with weight loss and diffuse lymphadenopathy, not focal. The mean age of onset is 50 years and it is slightly more common in men.

Knobs on RBC membranes and intrerythrocyte organisms, Choice C, describes the typical blood smear of a patient with malaria. Look for a history of recent travel in the patient history.

Microcytic, hypochromic red blood cells, Choice D, describes the typical smear for iron deficiency anemia. While possible in this case given the fatigue, it wouldn't explain that lymphadenopathy or sore throat.

Microspherocytes, Choice E, are a classic finding on a peripheral blood smear in a patient with hereditary spherocytosis. Look for a family history of anemia.
An otherwise healthy Kenyan man is prescribed trimethoprim-sulfamethoxazole for a urinary tract infection. He presents to the emergency department with jaundice, fatigue, and confusion. Which of the following might you see in the RBCs of a peripheral smear?

a. Heinz bodies  
b. Howell-Jolly bodies  
c. Basophilic stippling  
d. Auer rods  
e. Reed-sternberg bodies

**Correct answer:** A

G6PD deficiency is often asymptomatic until faced with an oxidative challenge, such as the drug TMP-SMX. It usually occurs in people of central African, Middle-Eastern, or Western Asian origin. The classic finding in RBCs is Heinz bodies (Answer A), which are collections of denatured hemoglobin. Howell-Jolly bodies are nuclear remnants in RBCs that remain due to failure of splenic sequestration in Sickle-cell disease. Basophilic stippling represents RNA collections that are most classically associated with lead poisoning. Auer rods are clumps of granular material diagnostic of acute myeloid leukemia (AML). Reed-sternberg bodies are the "owl-eyes" pathognomonic for Hodgkin’s lymphoma.
A 44 year old woman is brought by her husband to the local emergency department because she has "not been acting herself recently." The husband also reports that the patient has had a fever for the past 2 days, has been complaining of weakness and this morning noted that her urine was dark. Preliminary labs are taken that show:

- WBC: 13
- Hgb: 9 g/dl
- Platelets: 75,000
- Cr: 2.4
- BUN: 35
- PT: 12 (INR 1)
- PTT: 35

A blood smear is shown above.

What is the most likely cause for the patient's symptoms?

a. Disseminate intravascular coagulation
b. Hemolytic uremic syndrome
c. Hereditary spherocytosis
d. Idiopathic thrombocytopenic purpura
e. Thrombotic thrombocytopenic purpura

**Correct answer:** E

This patient is suffering from TTP (option E) which is a form of microangiopathic hemolysis that occurs when platelets aggregate in the small blood vessels. The platelet aggregation causes consumption of the platelets (thrombocytopenia) and leads to the hemolytic anemia in the form of schistocytes (picture) which will cause the dark urine. In addition, TTP is associated with renal failure, fever and change in mental status. This pentad Fever, Anemia (hemolytic with schistocytes), Thrombocytopenia, Renal Failure, Neuro (altered mental status) can be remembered with the mnemonic FAT-RN. Plasmaphoresis is the treatment of choice.

DIC (option A) is associated with schistocytes and hemolytic anemic however it is caused by consumption of coagulation factors and therefore one will see a rise in the PT and PTT which is normal in this case.

HUS (option B) presents similarly to TTP however it is usually the triad of anemia, renal failure and thrombocytopenia and NOT with fever and altered mental status. Usually HUS affects children and is associated with a bloody bacterial diarrhea usually caused by E. coli H7 O157 (EHEC).

Spherocytosis (option C) is also associated with hemolytic anemia with dark urine, but a blood smear would show round blood cells (spherocytes) and is not associated with renal failure, or mental status changes.

ITP (option D) is seen with a drop in platelets but not with hemolysis and renal failure.
which of the following protein is associated with band 3 of the rbc membrane protein?

a. actin
b. ankyrin
c. myosin
d. spectrin
e. glycophorin

**correct answer:** D
spectrin is attached to band 3 membrane protein of rbc.
A five-month old infant is brought to your office with a distressed mother. She complains that he bruises easily when on his back and arms when he is laid in his crib every night. She thinks his joints feel "loose" and that when she picks him up she is afraid that she may dislocate his shoulders. On exam his arms feature ecchymoses and cigarette-paper-like stretch marks on the skin. Which of the following is the likely defect causing this infant's presentation?

- a. Deficiency of sphingomyelinase
- b. Dynein arm defect
- c. Faulty collagen synthesis and misfolding
- d. Fibrillin gene mutation
- e. Glycogen accumulation in muscle tissue

**Correct answer:** C

This infant is exhibiting a classical form of Ehlers-Danlos syndrome, characterized by hyperextensible joints and skin under physiologic loading, and a tendency to bleed, due to a defect in collagen synthesis and folding.

Sphingomyelinase deficiency is associated with Niemann-Pick disease.

A dynein arm defect resulting in immotile cilia is found in Kartagener's syndrome.

The autosomal dominant fibrillin gene mutation is found Marfan's disease.

Glycogen accumulation in the muscle tissue of infants and toddlers can be verified microscopically in McArdle's disease, a disorder of glycogen storage.
A 65-year-old man presents with worsening symptoms of difficulty rising from a chair, shuffling gait, drooling, tremor at rest, and constipation. Which of the following is the most likely finding on examination of tissue if a brain biopsy was performed?

- a. Amyloid neuritic plaques in neocortex
- b. Atypical lymphocytes
- c. Eosinophilic sphere-shaped inclusions in cytoplasm
- d. Neurofibrillary tangles within neurons
- e. Protein aggregations and intranuclear inclusions

**Correct answer:** C

The correct answer is C, eosinophilic sphere-shaped inclusions in cytoplasm. These are Lewy bodies and they are the pathological hallmark of Parkinson’s disease along with degeneration of dopaminergic neurons of the substantia nigra. It is important to first recognize that this patient’s symptoms are seen in Parkinson’s disease.

The other histological findings are seen in other neurological diseases. Amyloid neuritic plaques and neurofibrillary tangles are seen in Alzheimer’s disease. Atypical lymphocytes may be seen in a viral encephalitis secondary to Epstein Barr Virus infection. Protein aggregations and intranuclear inclusions are findings in Huntington’s Chorea.
Over the course of one year, a previously healthy 45-year-old man with no significant family history developed worsening psychosis with neurologic degeneration and subsequently expired. Histological image of the brain is shown above.

Additional testing will most likely reveal which of the following findings?

- a. Abnormal tau proteins
- b. Expansion of DNA trinucleotide repeats
- c. Kernicterus
- d. Prions with beta-pleated sheet conformation
- e. Retrovirus proliferation

**Correct answer:** D

This patient suffered from variant Creutzfeld-Jacob Disease (vCJD) which is marked clinically by a rapid progressive dementia and histologically by spongiform changes in the cortex with loss of neurons. Though the exact mechanism of action is unclear, pathologic prions (as beta-pleated sheets, rather than alpha-helices) are seen (Choice D).

Abnormal tau (microtubule-associated) proteins (Choice A) are a hallmark of Alzheimer Disease, which manifests clinically as an insidious onset dementia with progressive memory loss and impaired executive functioning. The current patient is unlikely to have the disease, as incidence increases with age.

Expansion of trinucleotide repeats (Choice B) is the genetic basis of Huntington’s Disease. Though these patients may progress to dementia, onset is more gradual. Inheritance is autosomal dominant; lack of a family history also makes this choice unlikely.

Kernicterus (Choice C) is bilirubin deposits in the brain, usually basal ganglia, associated with hereditary hyperbilirubinemias and sulfonamide use in infants.

Retrovirus proliferation (Choice E) would be seen in HIV encephalopathy. This condition is unlikely given the present patient’s rapid course and prior medical history.
A pathologist is performing an autopsy on the brain of a 42-year-old woman who died in the same way her father and grandmother had which included a prolonged course of dementia, uncontrolled limb movements, and mood changes. What is the pathologist most likely to find on autopsy?

a. Cortical amyloid plaques  
b. Light colored substantia nigra  
c. Multiple ischemic lesions  
d. Shrunken head of the caudate  
e. Vacuoles and gliosis

**correct answer:** D

The age, genetic pattern and symptoms in the vignette suggest that the patient described likely suffered from Huntington's disease. The pathological findings characteristic of Huntington's include a shrunken head of the caudate (choice D) and dilated ventricles.

Cortical amyloid plaques (Choice A) are typical findings of Alzheimer's disease, but would be unlikely in a 42 year old and does not follow a clear genetic pattern.

Light colored substantia nigra (choice B) are classic findings in Parkinson's disease, where a loss of dopaminergic neurons occurs. Parkinson's is unlikely to cause dementia in such a young patient.

Multiple ischemic lesions (Choice C) are characteristic of multi-infarct dementia, which would be unusual in a woman of 42.

Vacuoles and gliosis (choice E) are typical findings in Creutzfeldt-Jakob Disease, which is generally not a genetic disease.
A medical student is examining slides of the urinary system under a microscope. She comes across a slide of stratified columnar epithelium. What part of the urinary system is this slide most likely to be from?

a. Female urethra  
b. Male urethra  
c. Renal pelvis  
d. Ureter  
e. Urinary bladder

**correct answer:** B

The only part of the urinary system that contains stratified columnar epithelium is the male urethra (Choice B), which can also contain pseudo-stratified columnar cells. The remainder of the urinary system contains transitional epithelium (Ureter - choice D, Bladder, Choice E, Renal Pelvis - Choice C) or stratified squamous (female urethra - choice A).
A 45 year old male presents to the Emergency Department of his local hospital coughing up blood. He reports a history of a dry cough for the past couple of months, but this is the first time that he has coughed up any blood. He denies any smoking history. On further questioning he notes that he has had episodes of blood in his urine recently.

A metabolic panel shows:
- Na: 140
- K: 4.9
- Cl: 105
- HCO3: 25
- BUN: 30
- Cr: 1.9

Urinalysis shows blood 2+, protein 1+, neg leukesterase, neg nitrites

Urine microscopy shows red blood cell casts

A kidney biopsy is taken and is stained (above). What is being stained in the slide?
- a. Anti-glomerular basement membrane Ab
- b. c-ANCA
- c. C3
- d. IgA
- e. p-ANCA

**correct answer:** A

This patient is suffering from Goodpasture's Syndrome which is an autoimmune disease caused by IgG attacking the Type IV collagen in the glomeruli and lung (option A Anti-GBM). Goodpasture's falls into the category of rapidly progressive glomerulonephritis (RPGN) which is the constellation of hematuria, red blood cell casts and acute renal failure. In cases of RPGN, a kidney biopsy is necessary which usually shows crescentic changes of the glomeruli on light microscopy and when stained for anti-GBM shows the clearly demarcated glomeruli basement membranes. Goodpasture's presents with kidney failure usually of the RPGN as well as hemoptysis although there may be only a dry cough on presentation. This is a serious condition that needs to be treated immediately with steroids and immunosuppressants.

c-ANCA (B) and p-ANCA (E) are seen as deposits in Wegener’s and Churg-Strauss nephritis, respectively. Wegener’s should be considered in RPGN but it usually presents with long history of sinus infections, and does not involve hemoptysis. Churg-Strauss vasculitis often has a history of asthma or other allergies. The ANCA stains of kidney biopsies show patchy deposits.

C3 deposits are seen in post-streptococcal glomerulonephritis. Therefore, there is usually a history of an upper respiratory infection or skin infection preceding the hematuria.

IgA is seen in IgA nephropathy (Berger’s Disease) which presents as hematuria presenting at the same time as a upper respiratory infection.
A 52-year-old female presents with chest pain associated with eating food. An endoscopic study shows ulceration in the lower esophagus. Histological examination of tissue from this area reveals mucosal ulceration, some blood, fibrin, and proliferation of blood vessels and fibroblasts. Many cells are undergoing mitoses, and most cells have prominent nucleoli. This histology most closely describes what process in this ulceration?

a. Caseating granulomatous inflammation  
b. Dysplastic epithelium  
c. Granulation tissue  
d. Noncaseating granulomatous inflammation  
e. Squamous cell carcinoma

**Correct answer:** C

A tissue undergoing repair requires regeneration of damaged cells and connective tissue replacement. Typically, granulation tissue (Answer C) is formed, characterized histologically by proliferating blood vessels and fibroblasts. Regenerating cells are characterized by mitoses and prominent nucleoli. This histological description should not be confused with dysplasia or malignancy.

Granulation tissue should not be confused with granuloma, which is a specific type of inflammation characterized by epithelioid cells, also known as activated macrophages.
Which of the following cells are responsible for producing a substance that reduces surface tension in the alveoli?

- a. Clara cells
- b. Macrophages
- c. Smooth muscle cells
- d. Type I pneumocytes
- e. Type II pneumocytes

**Correct answer:** E

Type II pneumocytes are responsible for producing surfactant, which reduces surface tension in the alveoli and prevents collapse of the lung.

(D) Type I pneumocytes are thin cells that comprise 97% of the alveolar lining. Their primary purpose is gas exchange. (A) Clara cells lack cilia and secrete glycosaminoglycans that are thought to protect the bronchiolar mucosa. (B) Macrophages and (C) smooth muscle cells do not secrete surfactant.
A 60 year old woman comes to your office for her follow-up visit after being hospitalized for an acute COPD exacerbation. She has a 30 pack year smoking history and complains of a chronic, productive cough. She is able to walk 50 feet before getting short of breath. On physical exam, there are decreased breath sounds bilaterally and crackles heard at both bases. It has been shown that cigarette smoking leads to goblet cell metaplasia and can contribute to the chronic bronchitis experienced by COPD patients. What level of the respiratory tract does NOT contain goblet cells?

a. Alveolar ducts  
b. Bronchi  
c. Bronchioles  
d. Respiratory bronchioles  
e. Terminal bronchioles

correct answer: E

The respiratory tract consists of many different types of cells. Pneumocytes, both type I and type II, are located in the alveoli. There is, however, different histology higher up in the respiratory tree.

Goblet cells exist in the bronchi and bronchioles (choices B and C), but do not extend into the terminal bronchioles (choice E). Goblet cells secrete mucus to trap foreign objects that will eventually be carried out of the lungs by the mucociliary system. Pseudocolumnar ciliated epithelial cells extend further, to the respiratory bronchioles. These cells move mucus secretions out of the lungs towards the mouth. (CONSIDER: As per high yield histology by Ronald Dudek, simple ciliated columnar epithelium starts at the level of Bronchioles. This transforms into simple ciliated cuboidal epithelium inside terminal and respiratory bronchioles)

Note: I think this question has more than one answer: goblet cells are not found in the alveolar ducts either.

Note 2: In that case they shouldn't be present in respiratory bronchioles either, apart from alveolar ducts. A, D, and E should all be correct.

Note 3: As per First Aid 2010, "goblet cells extend only to the terminal bronchioles" so only A and D are correct.
A 60 year old woman comes to your office for her follow-up visit after being hospitalized for an acute COPD exacerbation. She has a 30 pack year smoking history and complains of a chronic, productive cough. She is able to walk 50 feet before getting short of breath. On physical exam, there are decreased breath sounds bilaterally and crackles heard at both bases. It has been shown that cigarette smoking leads to goblet cell metaplasia and can contribute to the chronic bronchitis experienced by COPD patients. To what level do goblet cells extend in the respiratory tract?

a. Alveolar ducts
b. bronchi
c. bronchiole
d. respiratory bronchiole
e. terminal bronchiole

correct answer: E
A 48-year old female patient presents with a swelling in the neck. On examination, the patient appears cold-intolerant, bradycardic, and has a loss of the outer third of her eyebrows. She admits to have put on some weight in the past 2 years. Her blood results show raised antimicrosomal and antithyroglobulin antibodies. Which of the following correctly describes her condition?

a. It is associated with HLA-B27
b. It is associated with HLA-DR5
c. It is a type II hypersensitivity
d. It predisposes to medullary carcinoma of the thyroid
e. It is associated with insufficient iodine consumption

correct answer: B
Hashimoto’s thyroiditis is associated with HLA-DR5. It is a type IV hypersensitivity and predisposes to non-Hodgkin’s lymphoma.

HLA-B27 is associated with the seronegative spondyloarthropathies.

Insufficient iodine in the diet results in hypothyroidism and endemic goiter.
A child receives several vaccinations before the start of the school year. Shortly thereafter, he experiences a fever, joint pains, a rash and itching. The parents insist on a blood test, which reveals multinucleated giant cells with eosinophilic cytoplasmic and nuclear inclusion bodies. This is most likely a reaction to which vaccine?

a. Flu vaccine
b. Measles vaccine
c. Meningitis vaccine
d. Pneumococcal vaccine
e. Tetanus vaccine

correct answer: B

The live measles vaccine can cause mild measles symptoms, including multinucleated giant cells (a laboratory test for measles virus). Meningitis vaccine may cause pain and redness at the infection site. Tetanus vaccine may cause a type III allergic reaction at the infection site 24-48 hours after injection. Flu vaccine may cause flu-like symptoms. Some individuals may have a type I anaphylactic reaction to an egg protein used in the vaccine; persons with egg allergies should not get the flu vaccine. Pneumococcal vaccine may cause tenderness or redness at the injection site, fever, and joint aches.
A 38-year-old woman with history of Factor V Leiden deficiency, multiple deep venous thromboses, placement of IVC filter, and depression presents with syncope, severe pallor and abdominal distention after overdosing on her warfarin in a suicide attempt. Due to the severe bleeding, the patient requires fresh frozen plasma (FFP) to reverse the anticoagulation, but her blood type is unknown. Which of the following FFP types can be safely given?

a. A+
b. AB+
c. B-
d. O+
e. O-

**Correct answer:** B

Fresh frozen plasma (FFP) is indicated in severe bleeding to reverse anticoagulation. The woman has signs of hypovolemic shock and probable intra-abdominal bleeding that requires rapid correction. FFP contains clotting factors that are deficient in patients on warfarin (namely II, VII, IX, X). Plasma also contains antibodies to A and B antigens on RBCs. A and B antigens are glycosphingolipids. People with type A blood will produce antibodies to B antigen, people with type B blood will produce A antibodies, and people with type O blood will produce both. Similarly, people with Rh- type blood will produce antibodies to Rh factor. All these antibodies will be present in the plasma; therefore AB+ is the universal donor for PLASMA because it does not contain any ANTIBODIES. You may remember that O- is the universal donor for RBC because it does not contain any ANTIGENS. Incidentally, transfusion is not required to incite production of antibodies. Many foods, viruses, and bacteria can trigger sensitization to AB and Rh factors, so it is important to protect patients with unknown blood types by giving the universal donors.
In patients with AIDS, the failure to form granulomas in tissues infected by Mycobacterium tuberculosis is most likely due to which of the following?

a. Clonal deletion of T lymphocytes reactive to mycobacterial antigens
b. Decreased production of tumor necrosis factor-alpha by macrophages
c. Inability of T lymphocytes to provide helper functions for B lymphocytes
d. Inability of T lymphocytes to recruit and immobilize monocytes
e. Poor phagocytic activity of macrophages

**Correct answer:** D
A 54 year old man presents with severe pain in his back and in his ribs. Plain radiographs reveal several lytic bone lesions. Serum analysis reveals hypercalcemia and an M spike on serum protein electrophoresis. Urinalysis of this patient would reveal large amounts of which component of the protein shown below.

- a. I
- b. II
- c. III
- d. IV
- e. I + IV

correct answer: B

The correct answer is (B). Light chains of immunoglobulin G, known as Bence Jones Protein, are found on urinalysis of patients with multiple myeloma. This hematologic disorder consists of a triad of plasmacytosis (>10%), lytic bone lesions, and a serum M component. Plasma cell infiltration of the bone marrow leads to painful bone lesions and hypercalcemia due to osteoclast activation. Answer (A) contains the variable and CH1 regions of the heavy chain. Answer (C) labels variable regions of the heavy and light chains. Answer (D) refers to a portion of the heavy chain. Answer (E) are the parts comprising the heavy chain.
A 24-year-old man is involved in a motorcycle accident and loses several liters of blood. In addition to resuscitation with normal saline, physicians decide to transfuse blood products. If the patient has blood type B, Rh-negative, which of the following blood products may be most safely administered?

- a. Type A, Rh-negative red blood cells
- b. Type AB, Rh-negative red blood cells
- c. Type B, Rh-positive whole blood
- d. Type O, Rh-negative red blood cells
- e. Type O, Rh-negative whole blood

**Correct answer:** D

The key to this question is identifying what Rh and ABO blood group antigens and antibodies are present in the patient and in each blood product; a combination of antigen and antibody is considered an incompatibility, and will lead to hemolysis with possible renal injury or, in extreme cases, death. The patient expresses anti-A antibody, type B antigen, and anti-Rh antibody. Thus, Type O, Rh-negative red blood cells (Choice D) is the correct answer. They do not express type A, type B, or type Rh antigens, and have been removed from their donor plasma which originally contained anti-A, anti-B, and anti-Rh antibodies.

Type A, Rh-negative red blood cells (Choice A) is incorrect. Though this donor blood product does not contain any major blood group antibodies—red blood cells have been removed from donor plasma—donor RBCs will express both A antigen. The recipient’s anti-A antibodies will cause complement-mediated lysis of the donor RBCs due to expression of A antigen.

Type AB, Rh-negative red blood cells (Choice B) is incorrect. Though this donor blood product does not contain any major blood group antibodies—red blood cells have been removed from donor plasma—donor RBCs will express both A and B antigen. The recipient’s anti-A antibodies will cause complement-mediated lysis of the donor RBCs due to expression of A antigen.

Type B, Rh-positive whole blood (Choice C) is incorrect. Though the donor blood does not contain antibodies that are incompatible with the recipient, Rh antigen is expressed on the surface of donor RBCs. As the patient is Rh-negative, he has circulating anti-Rh antibodies that will cause complement-mediated lysis of the donor RBCs due to expression of Rh antigen.

Type O, Rh-negative whole blood (Choice E) is incorrect. Whole blood contains antibodies in the plasma component; thus Type O whole blood carries anti-A and anti-B antibodies. Though anti-A antibodies are compatible with this patient’s blood, anti-B antibodies are not compatible and will cause complement-mediated lysis of the recipient RBCs due to expression of type B antigen.
A 27-year-old African-American male with sickle cell anemia has had repeated bouts of infection with S. pneumoniae and N. meningitidis. How does his splenic dysfunction contribute to his immunocompromised state?

a. Decreased production of IgA
b. Decreased production of IgM
c. Deficiency in terminal complement factors
d. Impaired maturation of T cells
e. Impaired production of B cells

correct answer: B

When the spleen is dysfunctional (as in adult patients with SCA or post-splenectomy), presentation of antigen to B cells is impaired, leading to decreased production of IgM. Without the ability to activate complement to opsonize encapsulated bacteria, asplenic individuals are susceptible to infection by S. pneumoniae, H. influenzae, N. meningitidis, and Salmonella.

A -- IgA, the most common antibody in the body, is produced by B cells in the mucosa, not in the spleen.

C -- Deficiency in terminal complement factors prevents the formation of membrane attack complexes (MAC), leading to susceptibility to disseminated infection by Gram-negative bacteria such as Neisseria meningitidis. However, this deficiency has nothing to do with the spleen.

D -- T cells are necessary to activate B cells to produce antibodies, but T cells mature in the thymus, not the spleen.

E -- B cells are responsible for producing IgM in response to antigen presentation in the spleen. However, B cells are produced in the bone marrow, not the spleen.
A 17 year-old male is hiking through the foothills of West Virginia. Two days after his hike, he notices a large, inflamed region on his left calf characterized by redness, itching, and a rash. What type of hypersensitivity reaction is the young man experiencing?

- a. Type I anaphylactic
- b. Type II antibody-dependent
- c. Type III immune complex mediated
- d. Type IV delayed-type
- e. Type V stimulatory

**Correct answer:** D

The symptoms described are characteristic of poison ivy contact dermatitis. This is a Type IV Delayed-Type Hypersensitivity (DTH). It is mediated by sensitized T-cells that release cytokines when encountering the antigen.

When generally thinking about poison ivy and delayed type hypersensitivity (type IV) the rash would show up a few days later.
An 8-year-old boy presents to your clinic with a history of nonproductive cough and recurrent opportunistic infections. His mother claims that the symptoms first arose about 2 years ago and previous physicians suspected an immune deficiency disorder. A chest X-ray reveals patchy infiltrates and diffuse clouding. Bronchiolar lavage identifies numerous neutrophils arranged in isolated conglomerates. Nitroblue tetrazolium dye reduction test is negative. Which enzyme is most likely deficient in this child?

- a. Alpha-ketoglutarate dehydrogenase
- b. Dihydrofolate Reductase
- c. Glutathione Reductase
- d. NADPH Oxidase
- e. Topoisomerase

**Correct answer:** D

The child classically presents with chronic granulomatous disease, which results from NADPH Oxidase deficiency. Since NADPH Oxidase and related enzymes are critical in the phagocytic responses of neutrophils, the deficient neutrophils are incapable of destroying common bacteria, and opportunistic infections abound. The nitroblue test is specifically performed to determine the presence of NADPH Oxidase.
An otherwise healthy 10-year-old boy develops blotchy areas of erythema that are pruritic over the skin of his arms, legs, and trunk within an hour every time he eats seafood, followed by diarrhea. These problems abate within a few hours, and physical examination reveals no abnormal findings. Which of the following immunologic mechanisms is at work?

b. Hypergammaglobulinemia
c. Immune complex deposition
d. Localized anaphylaxis
e. Release of complements

**correct answer:** D
This is a classical type I hypersensitivity reaction (food allergy).

Cell mediated hypersensitivity is a Type IV.
Hypergammaglobulinemia is plasma cell mediated.
Complement C3B is a Type III.
Immune complex deposition is also Type III.
An 80 year-old woman with metastatic vulvar carcinoma has marked temporal wasting and has lost 30 lbs over the last 2 months. Which cytokine is thought to contribute to the cachexia seen in advanced cancer?

- a. APC
- b. CEA
- c. IL-4
- d. IL-5
- e. TNF-alpha

**Correct answer: E**

TNF-alpha, also called cachectin, is an acute phase reactant secreted mainly by macrophages. Chronically elevated levels of TNF-alpha cause cachexia, which is characterized by weight loss, muscle atrophy (usually most noticeable in the temporal region), weakness, and anorexia. It is also important to remember that septic shock is also mediated through TNF-alpha.
A 22-year-old woman develops diarrhea with foul smelling, floating stools for several months. She undergoes an endoscopy with biopsy of the small intestine demonstrating villous atrophy. She now presents to the physician complaining of a new rash. On exam, she has a pruritic, papulovesicular rash covering her trunk. The doctor reassures her that the recommended treatment for her gastrointestinal disorder will help to resolve the rash as well. If this patient were to undergo a skin biopsy, what class of antibody would likely be present?

a. IgA
b. IgD
c. IgE
d. IgG
e. IgM

**Correct answer:** A

This patient has celiac disease, or gluten-sensitive enteropathy, and dermatitis herpetiformis. Celiac disease results from reaction to the wheat protein gliadin resulting in autoimmune attack on the small bowel. Classic celiac disease is defined by three features: (1) villous atrophy (2) symptoms of malabsorption such as steatorrhea, weight loss or other nutrient deficiency and (3) resolution of the mucosal lesions upon removal of gluten-containing products. Serologic studies reveal IgA autoantibodies to gliadin and to proteins in the small bowel that are the target of the disease.

Dermatitis herpetiformis is a condition seen commonly in patients with celiac disease. It is characterized by pruritic papulovesicles on the trunk and the arms. On biopsy, characteristic granular deposits of IgA on the basement membrane are seen.

B. IgD is primarily found on the surface of B-cells.

C. IgE plays an important role in the response to parasites and in allergic reactions.

D. IgG is the most abundant immunoglobulin making up 75% of the serum immunoglobulins. IgG is the only antibody that can pass through the placenta. IgG plays a role in the immune reactions against many pathogens including bacteria, viruses and fungi.

E. IgM is the largest immunoglobulin and the basic immunoglobulin present on B-cells. IgM is secreted early in an infection so the presence of IgM can help determine the time course of the infection.
A 32-year-old African American female presents to your clinic with a temperature of 104 deg Fahrenheit and generalized fatigue and lethargy. The cytokine most likely responsible for her increased temperature is secreted by which of the following cells?

a. B cells
b. Fibroblasts
c. Macrophages
d. Neutrophils
e. T cells

correct answer: C

Interleukin-1 (IL-1) is the major endogenous pyrogen and is responsible for the generation of most fever symptoms. IL-1, secreted by macrophages, serves to stimulate T cells, B cells, neutrophils, fibroblasts and epithelial cells to grow, differentiate and synthesize specific products.

You should be familiar with the functions of IL-1 through 5. The mnemonic HOT T-BONE stEAk may help:

- IL-1 – HOT (fever)
- IL-2 stimulates T cells
- IL-3 stimulates bone marrow
- IL-4 stimulates IgE production
- IL-5 stimulates IgA production

B cells (A) are involved in antibody mediated immunity and may mature into plasma cells that secrete IgA, G, and E antibodies.

Fibroblasts (B) performs multiple functions as they are undifferentiated precursors to a variety of cell types including epithelial cells.

Neutrophils (D) secrete various cytokines but are mainly involved in phagocytosis of bacterial organisms.

T cells (E) are derived from Th1 helper cells. Secretion of gamma interferon by Th1 cells activates macrophages, while IL-2 secretion by Th1 cells activates cytotoxic cells. Native helper T cells (Th0) are induced to form Th2 cells by IL-4. Th2 secretion of IL-4 and IL-5, in turn, induce B cell conversion to plasma cells.
A 28-year-old man presents to the emergency room complaining of shortness of breath and coughing up bright red blood. He reports several months of dry cough and fatigue. On examination, blood pressure is 150/98 mmHg, and respiratory rate is 28/min. Laboratory examination is remarkable for a BUN of 30 mg/dL and creatinine of 2.9 mg/dL. Kidney biopsy confirms the diagnosis.

**Which of the following human leukocyte antigens (HLA) is most strongly associated with this patient’s disease?**

a. HLA-DR2  
b. HLA-DR3  
c. HLA-DR4  
d. HLA-DR5  
e. HLA-DR7

**Correct answer:** A

This patient is suffering from Goodpasture's syndrome, characterized by pulmonary hemorrhage and glomerulonephritis. This disease is associated with major histocompatibility complex (MHC) class II antigen HLA-DR2. The DR2 antigen is also associated with allergy, multiple sclerosis, and narcolepsy. The underlying pathology in this disorder is an autoimmune attack against basement membrane antigens—it is also known as anti-glomerular basement membrane disease—explaining the often-simultaneous involvement of the lungs and kidneys. Kidney biopsy with immunohistochemical staining provides diagnosis.

HLA-DR3 (choice B) is associated with celiac disease, insulin-dependent diabetes, and systemic lupus erythematosus.

HLA-DR4 (choice C) is associated with pemphigus vulgaris, rheumatoid arthritis (relative risk 4), and insulin-dependent diabetes mellitus.

HLA-DR5 (choice D) is associated with juvenile rheumatoid arthritis and pernicious anemia.

HLA-DR7 (choice E) is associated with psoriasis.
Two patients are vaccinated against poliomyelitis. One patient receives the killed vaccine and the other patient receives the live attenuated vaccine. One month after the vaccination, the levels of which of the following poliovirus antibodies differ most between these patients?

a. Serum IgM
b. Serum IgG
c. Serum IgA
d. Cerebrospinal fluid IgG
e. Duodenal IgA

**Correct answer:** E

One general principal of vaccination dictates that local secretory antibodies synthesizes is best promoted when specific mucosal surfaces are directly stimulated by antigen. When both live and killed vaccines are applied to a mucosal surface, the live attenuated viral vaccines viral vaccines appear more affected of the two in generating prolonged mucosal IgA secretion. The live vaccines are thought to colonize the natural site of viral entry, producing a greater and more prolonged immune response there. Killed viral vaccines (which are usually administrated intramuscularly) are unable to stimulate mucosal secretory IgA production. In case of the polio vaccines, the intramuscularly given Salk formaldehde inactivated polio vaccine (IPV) is less effective in generating a prolonged nasopharyngeal and intestinal mucosal secretory IgA response than is the Sabin live attenuated oral polio vaccine (OPV).

(CHOICES A, B, D) The antibody most strongly associated with antigenic mucosal contact is IgA, NOT IgG or IgM.
(CHOICE C) The two sites of primary replication (and associated mucosa, IgA secretion) are the oropharynx and the intestine. The serum IgA levels appear less impacted by vaccinated
This immunoglobulin is the second to respond to an antigen. It has a half life of 6 days.

- a. IgA
- b. IgG
- c. IgD
- d. IgE
- e. IgM

The correct answer is A. The Correct answer is IgA, it is a dimer, predominant in saliva and other secretions with a half life of six days. The first to attack is IgM then IgA followed by IgG.
MCH-II presents on these cells except:

a. Macrophages
b. Dendritic Cells
c. Somatic Cells
d. B lymphocyte Cells
e. Antigen Presenting Cells (APC) Cells

correct answer: C
Somatic Cells Presents MCH-I Only....
A 28-year-old man presents to the emergency room complaining of shortness of breath and coughing up bright red blood. He reports several months of dry cough and fatigue. On examination, blood pressure is 150/98 mmHg, and respiratory rate is 28/min. Laboratory examination is remarkable for a BUN of 30 mg/dL and creatinine of 2.9 mg/dL. Kidney biopsy confirms the diagnosis.

Which of the following human leukocyte antigens (HLA) is most strongly associated with this patient’s disease?

a. HLA DR2
b. HLA DR3
c. HLA DR4
d. HLA DR5
e. HLA DR7

**Correct answer:** A

Choice E is HLA DR7 which is associated with steroid responsive nephrotic syndrome and psoriasis is associated with HLA B27.
A 40 year old female presents to her physician for worsening joint pain. She has had joint pain for many years in both wrists, MCP joints, and PIP joints. Her DIPs are spared. Her pain is always worse in the morning and improves as the day goes on. Lately she realizes both knees also hurt her and she is more tired than usual. Examination reveals her painful joints to be mildly warm and her PIPs are flexed, while her DIPs are extended. Laboratory testing reveals high titers of rheumatoid factor (RF), an elevated ESR and C-reactive protein, and a normocytic normochromatic anemia. Examination of the synovial fluid at the involved joints would most likely reveal which of the following?

- Clear fluid; 125 WBC/mm³; 20% PMNs
- Clear-yellow fluid; 1,800 WBC/mm³; 20% PMNs
- Cloudy-yellow fluid; 6,000 WBC/mm³; 60% PMNs
- Turbid, purulent fluid; 50,000 WBC/mm³; 80% PMNs
- Reddish fluid; 1,500 WBC/mm³; 20% PMNs

**Correct answer:** C

The correct answer is C.

The patient described likely has rheumatoid arthritis. This chronic inflammatory autoimmune disease involves the synovium of the joints, causing damage to the cartilage and bone. Women are more commonly affected and HLA-DR4 is associated. High RF is associated with severe disease, although 3% of the healthy population has RF. RA exhibits symmetrical arthritic of proximal joints (wrists, MCP, PIP, elbow, knee) often worse in the morning. DIP are not affected. The patient here also exhibits Boutonniere deformities, commonly seen with RA. Synovial analysis will show findings consistent with an inflammatory process (clear/yellow fluid; <2000 WBC; 50-70% PMNs). Only choice C fits these criteria.

Choice A would be the result from a healthy, normal joint, (clear fluid; <200 WBC; <25% PMNs).

Choice B would be seen in an osteoarthritis joint, (clear-yellow fluid; <2,000 WBC; <25% PMNs).

Choice D would be seen in a septic joint, (turbid fluid; >50,000 WBC, >70% PMNs) either bacterial or tuberculosis. Septic arthritis requires urgent treatment as permanent joint destruction can result in as little as 24 hours.

Choice E would be seen in a joint that had experienced trauma, (red secondary to RBC; <2,000 WBC; <25% PMNs).

***I do not think that choice D could describe septic arthritis with Tuberculosis. In this case you would have a higher lymphocyte percent than PMN's.
A 55 year old woman presents to her physician with complaints of bilateral joint pain in her wrists, hands, and knees, muscle aches, fatigue, and malaise for the past two months. Her past medical history includes uncontrolled hypertension, for which she was prescribed hydralazine 3 months ago. Laboratory testing reveals a positive ANA, however anti-DNA antibodies are negative. Which of the following antibodies would be most specific to her disease?

a. Antihistone
b. Anticentromere
c. Antiscleroderma-70
d. Anti-Sm
e. Anti-uroporphyrin isomerase ribonucleoprotein

**Correct answer: A**
Choice A (antihistone) is correct.

This patient likely has drug-induced lupus from hydralazine. Other drugs causing this syndrome include procainamide, isoniazid, chlorpromazine, methyldopa, and quinidine. Most patients improve after removal of the offending agent. Drug induced lupus is similar to SLE, except there is no CNS or renal involvement. Also, malar rash, alopecia, and ulcers are typically not seen. Antihistone antibodies are always present and there is an absence of anti-ds DNA and anti-Sm Ab.

Choice B (anticentromere) is likely to be seen in limited scleroderma (CREST).

Choice C (antiscleroderma-70) is very specific for diffuse scleroderma but is not always present. It is also known as antitopoisoamerase I.

Choice D (anti-Sm) is most likely to be seen in systemic lupus erythematosus.

Choice E (Anti-uroporphyrin isomerase ribonucleoprotein) is most likely to be seen in mixed connective tissue disease in which symptoms of SLE, RA, scleroderma, and polymyositis.
A 38 year old woman with a 15 year history of drug abuse who is being treated for fever, cough, and shortness of breath, develops dementia, hemiparesis, ataxia, aphasia, and dysarthria over the course of 5 days. MRI reveals multiple white matter lesions. Over the course of 2 weeks, the woman's condition worsens, despite aggressive treatment, she lapses into a coma and soon dies. At autopsy, histologic examination of her brain tissue reveals gigantic, deformed astrocytes and oligodendrocytes with abnormal nuclei.

What is the most likely cause of this woman's CNS symptoms?

a. Autoimmune attack of myelin sheaths
b. A double-stranded circular DNA virus
c. A single-stranded linear RNA virus
d. An autosomal recessive lysosomal storage disease
e. A proteinaceous infectious particle

Correct answer: B

The correct answer is (B). JC virus is a ubiquitous polyomavirus that causes progressive multifocal leukoencephalopathy (PML). PML is associated with severely immunocompromised states. Most cases are seen in patients with AIDS. The disease can have a very rapid course, progressing over days or weeks. A CD4 count less than 100 is an especially poor prognostic sign in these patients. Characteristic histologic findings include giant astrocytes, and oligodendrocytes with enlarged nuclei and nuclear inclusions. Radiographic findings include numerous white matter lesions of varying sizes in the cerebral and cerebellar hemispheres. Answer (A) alludes to multiple sclerosis a demyelinating disease defined by multiple white matter lesions separated over time and space. Answer (C) refers to HIV encephalitis, which progresses slowly over a course of months. Answer (D) refers to metachromatic leukodystrophy, an autosomal recessive lysosomal storage disease that causes both peripheral and central demyelination in children. Answer (E) refers to prion disease, which would reveal spongiform encephalopathy.
Two weeks after a 4 day bout of gastroenteritis, a man develops burning sensations in his toes. The following day, he experiences weakness in both legs. The weakness in his legs progresses to involve his arms 3 days later. On examination, you note facial flushing, systolic blood pressures ranging from 70 to 160, profuse diaphoresis, and bilateral facial paralysis. Lumbar puncture reveals a normal white cell count and elevated protein. He is treated with plasma exchange and intravenous immunoglobulin and steadily recovers completely in 3 months. The type of hypersensitivity reaction that caused this patient’s condition is most similarly described by which of the following:

a. A 2nd year medical student returns to the student health center 48 hours after PPD placement and is told that her PPD test is positive
b. Within minutes of anastamosis, a transplanted kidney becomes cyanotic and mottled
c. A 43 year old obese woman with a fasting blood sugar of 220 mg/dL begins treatment with metformin
d. Moments after being stung by a bee, a young girl begins wheezing and develops hives
e. Three days after taking penicillin, a patient develops fever, hives, arthralgias, lymphadenopathy, and eosinophilia

Correct answer: A

The correct answer is (A). A PPD test relies on the presence of previously sensitized CD4+ lymphocytes in individuals infected with M. Tuberculosis. This is a type of type IV hypersensitivity reaction. The question stem describes findings of Guillain-Barre Syndrome, another type IV hypersensitivity reaction caused by a T cell mediated, mononuclear inflammatory infiltration of nerves and nerve roots accompanied by destruction of myelin.

Answer (B) describes hyperacute transplant rejection, a type II hypersensitivity reaction caused by preformed antibodies against foreign antigens on transplant tissues.

Answer (C) describes type II diabetes, which is not associated with hypersensitivity reactions. However, be aware that type I diabetes is caused by a type IV hypersensitivity reaction.

Answer (D) describes anaphylaxis, a type I hypersensitivity reaction.

Answer (E) describes serum sickness, a type III hypersensitivity reaction. Serum sickness is most commonly caused by penicillin. It is treated with antihistamines and corticosteroids.
A 42-year-old male presents to you with recent episodes of bloody urine and blood streaked sputum. A biopsy of his kidney is taken and immunofluorescence demonstrates linear deposits on the basement membrane. The reaction which led to the patient’s condition is most similar to which of the following reactions?

a. Bee sting
b. Henoch–Schönlein purpura
c. Poison ivy
d. Positive PPD test
e. Graves’ disease

correct answer: E

Hemoptysis and hematuria are two of the presenting symptoms of Goodpasture’s syndrome, which is this patient’s diagnosis. This condition develops via a type II hypersensitivity reaction with antibodies to the glomerular basement membrane. The finding of linear deposits on immunofluorescence helps to confirm the diagnosis. Of the choices listed, only Graves’ disease is a type II hypersensitivity reaction. Contact dermatitis due to poison ivy and a positive PPD test are examples of type IV delayed hypersensitivity. Henoch–Schönlein purpura is an immune complex type III hypersensitivity with IgA deposits in blood vessels. The reaction to a bee sting would be an example of an anaphylactic reaction, which is a type I hypersensitivity.
A 67-year-old woman with long standing rheumatoid arthritis has been treated with gold for a number of years. On a routine examination, edema, 4+ protein in the urine with lipiduria, and a serum albumin of 2.5 gm/dL are noted. A renal biopsy is performed and on light microscopy marked diffuse thickening of the glomerular capillary walls are noted. Which of the following mechanisms is most likely responsible for causing the glomerular capillary walls to become leaky, leading to this woman's nephrotic syndrome?

a. Altered renal tubular epithelial with cyst formation
b. Chronic activation of C5b-C9 pathway on glomerular endothelial and mesangial cells
c. Hypoperfusion of the glomerulus
d. Ischemic damage to the tubular epithelium leading to necrosis
e. Persistent overexpression of ENaC in cortical collecting tubule

**correct answer:** B

Answer is B: This woman has membranous glomerulopathy. In 85% of patients this disease is considered idiopathic, but it has been associated with drugs such captopril, NSAIDs, penicillamine and gold. Experimental evidence suggests that capillary wall injury is mediated by chronic activation of C5b-C9 pathway on glomerular endothelial and mesangial cells which leads to formation of the MAC complex. This induces epithelial and mesangial cells to liberate proteases and oxidants, damaging the capillary walls and making them leaky.

Choice A is seen in in polycystic kidney disease, but is not associated with membranous glomerulopathy.

Choice C is thought to be the cause of temporary kidney dysfunction seen in pre-renal azotemia.

Choice D is the cause of dysfunction in acute tubular necrosis (ATN).

Choice E is seen in a Liddle's Syndrome.

A 4-year old boy presents with a fever, coughing and sneezing for the last 3 days. The patient’s parents report that he has suffered severe upper respiratory and sinus infections since birth. Physical exam reveals inflammation and pus in the sinuses. A bacterial culture indicates Haemophilus influenzae infection. A panel of genetic tests is ordered, which shows a hereditary immune deficiency which affects the complement cascade. Which of the following is most likely to be deficient in this patient?

a. C1 Esterase  
b. C3  
c. C6  
d. C8  
e. DAF

**correct answer: B**

This is a classical description of C3 deficiency, which leads to severe pyogenic sinus and respiratory infections in childhood, especially with encapsulated bacteria such as Haemophilus influenzae.

C1 esterase deficiency causes hereditary angioedema.

C6 and C8 deficiency causes Neisseria infections.

DAF deficiency causes paroxysmal nocturnal hemoglobinuria.
A 41-year-old woman suffers from dysphagia, crops of red skin spots, and painful blue hands when exposed to cold. She is started on monotherapy for latent tuberculosis but immediately develops a malar rash. Which of the following antibody tests could be positive in this woman?

a. Anti-scl 70  
b. Anti-centromere  
c. Anti-microsomal  
d. Anti-smooth muscle  
e. Anti-Ro

**Correct answer: B**

The woman’s disease described includes Esophageal dysmotility, Telangiectasias, and Raynaud’s phenomenon of CREST syndrome, or limited scleroderma. The other features are calcinosis and sclerodactyly. Anti-centromere antibodies are associated with CREST.

The second part of the question describes drug-induced lupus from isoniazid. Classic drug causes include hydralazine, procainamide > phenytoin and amiodarone. Anti-histone antibodies (Answer B) are associated.

Anti-scl 70 antibody is associated with diffuse scleroderma, which does not exhibit CREST syndrome.

Antimicrosomal antibody is associated with Hashimoto’s thyroiditis.

Anti-smooth muscle antibody is associated with autoimmune hepatitis.

Anti-Ro antibody is associated with Sjogren’s syndrome.

Additionally, it is possible that this woman is having symptoms of mixed connective tissue disease, which can have SLE symptoms and is associated with anti U1-RNP antibody. She may also have SLE alone, which would be associated with ANA and anti ds DNA or anti-Smith antibody.
which of the following has subepidermal blisters?
   a. dermetitis hepertiform
   b. bullous pemphigoid
   c. leprosy
   d. scabies
   e. pemphigus vulgaris

**correct answer:** B
this is the disease which u always differentiate from p. vulgaris
A one-month-old infant with no past medical history, normal gestation and delivery is brought to the hospital in cardiogenic shock. A viral infection is suspected for the acute heart failure. Which of the following viruses is the most likely culprit?

- a. Coxackie Virus
- b. Herpes Simplex Virus 2
- c. Respiratory Syncytial Virus
- d. Rotavirus
- e. Rubella

**Correct answer:** A

Coxackie Virus B causes can cause myocarditis and cardiogenic shock from acute heart failure. Infants may have been exposed to sick contacts with mild viral syndromes. Herpes Simplex Virus (HSV) 1 or 2 can cause severe perinatal complications including seizures, meningitis and sepsis, and may cause neurogenic shock but heart failure is not usually the presenting scenario, and 1 month is not perinatal. Maternal rubella infection can also cause severe congenital defects but has a mild course when the infection is not perinatal. Rotavirus can cause severe gastroenteritis in children, leading to hypovolemic shock from diarrhea and vomiting. Respiratory Syncytial Virus (RSV) causes bronchiolitis in infants.
A 43-year-old female presents to your office complaining of shortness of breath when she walks briskly. She also reports waking up in the middle of the night short of breath four to five times a week for the past eight months. She is a recent immigrant from Thailand where she was a sex worker for the past 15 years. Physical exam is significant for a high-pitched diastolic murmur, bounding pulses in the upper extremities, rales in the lower lung lobes and 2+ pedal edema. It is noted that she is rhythmically bobbing her head during the exam. Which of the following is the likely mechanism for her condition?

- a. Airborne transmission
- b. Congenital malformation
- c. Drug abuse
- d. Oncogenesis
- e. Sexual transmission

**correct answer:** E

This patient is presenting with likely syphilitic aortitis, a condition classically seen in the tertiary phase of the disease. It can present insidiously without earlier signs of the syphilis, and is usually contracted 1-10 years prior via sexual contact with an individual with open primary syphilis lesions. In particular, the patient's social history as a sex worker was indicative given the signs of aortic regurgitation and heart failure. Additionally, de Musset's sign of head bobbing is a sign of aortic regurgitation.
A 52-year-old male immigrant from Bolivia presents to the clinic complaining of dyspnea, weakness, and mild edema. Cardiac ultrasound reveals dilatation and hypertrophy of all four chambers. Myocardial biopsy reveals inflammation with an eosinophilic infiltrate.

What is the most likely etiologic agent for this man’s condition?

a. Coccidioides immitis  
   b. Coxsackievirus A  
   c. Naegleria fowleri  
   d. Streptococcus viridans  
   e. Trypanosoma cruzi

**correct answer: E**

This patient's history of congestive heart failure symptoms and imaging study are consistent with a diagnosis of dilated cardiomyopathy. Biopsy result indicate a protozoal causative agent; Trypanosoma cruzi (choice E), causative agent of Chagas disease, is consistent with the given information including country of origin.

Coccidioides immitis (choice A) is endemic to parts of South America, but it more typically manifests with respiratory symptoms.

Coxsackievirus A (choice B) is the causative agent in hand, foot and mouth disease. (Coxsackievirus B is the strain commonly seen in myocarditis.) Additionally, biopsy of a viral myocarditis would be associated with a mononuclear infiltrate, not eosinophilic as seen here.

Naegleria fowleri (choice C) is the causative agent in primary amebic meningoencephalitis. It does not cause myocarditis.

Streptococcus viridans (choice D) is seen primarily in subacute bacterial endocarditis, not myocarditis. Common agents in bacterial myocarditis include diptheria and meningococci. Additionally, biopsy of a bacterial myocarditis would be associated with a neutrophilic infiltrate, not eosinophilic as seen here.
A 48 year-old female presents with progressive shortness of breath and anxiety. She has no notable prior medical history, and on physical exam, auscultation reveals an opening snap over the cardiac apex followed by a mid-diastolic rumble. The most common cause of her disease process is which of the following?

- a. chronic hypertension causing dilatation of the left ventricle
- b. repeated attacks of Streptococci causing valvular lesions
- c. primary pulmonary hypertension causing atrial dilatation
- d. a congenital atrial-septal defect causing chronic hypoxemia
- e. a congenital bicuspid valve causing increased afterload of the left ventricle

**Correct answer**: B

The answer is B. The patient has mitral valve stenosis. Repeated attacks of group A Streptococci may cause rheumatic fever, which is an auto-immune disease (type II hypersensitivity) where antibodies to the bacteria cross-react with valvular proteins. This is referred to as molecular mimicry. Most commonly the mitral valve is affected, and the aortic valve is the second most commonly involved valve.

Choice A is incorrect; chronic hypertension may indeed cause left ventricular dilatation or more classically left ventricular hypertrophy. This may ultimately result in mitral regurgitation and congestive heart failure.

Choice C is incorrect; primary pulmonary hypertension causes right sided heart failure and signs consistent with tricuspid valvular regurgitation.

Choice D is incorrect; an atrial septal defect should not cause mitral valve stenosis. About half of ASD's correct themselves without surgery by age 5 years.

Choice E is incorrect; the mitral valve normally has two leaflets. A congenital bicuspid aortic valve is more prone to calcification and lead to aortic valvular stenosis at an earlier age. Left ventricular hypertrophy may result with a late-peaking holosystolic crescendo-decrescendo murmur best heard over the right sternal border.
A 56-year-old woman has a long-standing heart murmur. You hear an opening snap followed by a mid-diastolic rumble. Type II hypersensitivity to a virulence factor caused this patient’s valvular disease. What is the pathogenic activity of this virulence factor?

a. ADP-ribosylates Gi thereby increasing cAMP
b. Activates T cells to release IFN-gamma and IL-2
c. Binds with factor H to decrease complement activation
d. Causes an exfoliative rash
e. Inactivates elongation factor 2 (EF-2)

**Correct answer:** C

The vignette describes the murmur of mitral stenosis, a common valvular disease seen in chronic rheumatic fever. RF begins with an upper respiratory Streptococcus pyogenes infection. Then a type II hypersensitivity (ie. autoimmune) reaction to the M protein damages the heart and valves. M protein binds with factor H to decrease complement activation (C).

A -- The toxin of Bordetella pertussis ADP-ribosylates Gi, inactivating it and thereby increasing cAMP. This leads to the characteristic whooping cough.

B -- Staphylococcus aureus makes Toxic Shock Syndrome Toxin-1, a superantigen that activates T cells to release IFN-gamma and IL-2.

D -- Staphylococcus aureus also makes exfoliative toxins A and B, which cause Scalded-Skin Syndrome, a condition seen most often in infants that looks worse than it really is.

E -- Corynebacterium diphtheriae toxin ADP-ribosylates elongation factor 2 (EF-2). This interrupts protein synthesis, which kills the host cells and creates a mat of debris (pseudomembrane) in the oropharynx.
A 54 year old male with a history of chronic liver disease presents to your clinic for the first time. After conducting a full history, you perform the physical exam and notice gynecomastia, a protruding abdomen, testicular atrophy, and several spider angiomas. Which of the following represent(s) the possible underlying cause of his disease?

a. Hepatitis A infection
b. Hepatitis B infection
c. Hepatitis C infection
d. A and B only.
e. B and C only.

correct answer: E

Hepatitis B and C infection can both result in chronic liver disease marked by certain tell tale signs in the H&P. Portal hypertension leads to ascites, splenomegaly, caput medusa, esophageal varices, hemorrhoids. An increase in estrogen-related hormones leads to gynecomastia. Spider angiomas are also common in these patients. HBV is acquired primarily through sexual contact and materfal-fetal transmission. HCV is acquired primarily through parenteral modes such as IV drug use and blood transfusions. Sexual transmission of HCV has also been reported, but is a less common cause.

Hepatitis A causes an acute hepatitis usually marked by diarrhea, jaundice, fever, and RUQ pain. The infection is typically cleared shortly thereafter and does not lead to portal hypertension.
A 4-year-old boy presents to the emergency room after two days of steadily worsening right lower quadrant pain and occasionally bloody diarrhea. On careful questioning, the patient’s father thinks that there was another child in his day care center that had a similar condition. Physical exam does not show rebound tenderness but a CT exam shows signs of diffuse lymphadenitis. The patient’s father is nervous because he has ulcerative colitis and is wondering if his son is developing it as well. What is the most likely cause of this patient’s condition?

- a. Appendicitis
- b. E. coli O157:H7 infection
- c. Flare of inflammatory bowel disease
- d. Shigella infection
- e. Yersinia infection

**Correct answer:** E

This patient likely has an infection of *Yersinia enterocolitica*. It frequently presents with bloody diarrhea and right lower quadrant pain which can mimic Crohn's disease and appendicitis. Peritoneal signs are not present, which can help distinguish it from appendicitis. In some patients, mesenteric lymphadenitis is present. Furthermore, in some patients with the HLA-B27 serotype, this infection can result in seronegative arthritis.

While both E. coli O157:H7 and shigella infection can result in bloody diarrhea, they would not normally present with diffuse lymphadenopathy.
Eight patients enter your office on a hot July afternoon. The patients are all attending a family reunion. This morning the family ate eggs and toast. For lunch they had potato salad and fried chicken. All eight of them complain of nausea, vomiting, and stomach aches. They are concerned about Celiacs since that runs in their family. What is the most probable agent responsible for their symptoms?

a. An antigliadin antibody
b. A gram positive, catalase positive bacteria
c. A gram positive, catalase negative bacteria
d. A toxin produced by a gram positive, catalase positive bacteria
e. A gram negative rod

**Correct answer:** D

S. aureus is the most common cause of food poisoning, however the preformed toxin that S. aureus produces is the primary agent by which food poisoning is caused.

A- It is unlikely that the family would all experience previously unknown symptoms for celiacs disease at the same time.

B- This is a good answer since S. aureus is a Gram positive catalase positive organism. However the bacteria itself is not the cause of the patients symptoms. Answer choice D is more correct.

C- S. aureus is catalase positive. This answer would describe Streptococci or Enterococci. Both are unlikely to cause food poisoning.

E- Infections involving bacteria take some time before showing some effect. Considering this to be C. jejuni it would take some time before symptoms arose from the infection. Since the family has had only two meals together the time frame doesn’t match. Also, infection caused by C. jejuni is less common than food poisoning.
To which receptor on B Cells does the Epstein-Barr Virus attach?

a. CD19
b. CD20
c. CD21
d. CD4
e. CD8

**correct answer:** C

The Epstein-Barr Virus attaches to CD21 on B cells.
A 19-year-old man has a 1-week history of abdominal cramps and bloody diarrhea. His temperature is 102 degrees Fahrenheit, and there are rose spots on his abdomen. Which of the following is most likely to be a characteristic of the causative organism?

- a. Actin polymerization
- b. Lactase-positive
- c. Nonmotile
- d. Oxidase-positive
- e. Produces H2S

**Correct answer: E**

This patient has typhoid fever (dysentery, fever, rose spots), a systemic, human-specific infection that manifests with gastroenteritis 1-2 weeks after initial infection and takes 4-6 weeks to resolve. The causative organism, Salmonella typhi, is a Gram-negative, flagellated rod that ferments glucose but not lactose. It is oxidase-negative and produces H2S. It does not polymerize actin.

**A --** Actin polymerization is a feature of both Listeria monocytogenes and Shigella. Listeria may cause mild gastroenteritis in healthy individuals, while Shigella is a virulent cause of invasive dysentery.

**B --** Enteric pathogens that can metabolize lactose include E. coli and Klebsiella. Many strains of E. coli cause gastroenteritis: EIEC causes invasive dysentery, EHEC causes non-invasive dysentery, ETEC causes traveler's diarrhea, and EPEC causes diarrhea mostly in children. Although it is part of the intestinal flora, Klebsiella is primarily pathogenic when aspirated, leading to pneumonia or lung abscesses.

**C --** Nonmotile enteric pathogens include Klebsiella and Shigella.

**D --** Oxidase-positive enteric pathogens include Vibrio cholerae and Campylobacter. The 1A:5B exotoxin of cholera ADP-ribosylates Gs, increasing cAMP and causing secretory diarrhea ("rice water stools"). Campylobacter colonizes the mucosa of the small intestine and causes watery or bloody diarrhea.
A 46-year-old man goes on a camping trip and ingests undercooked pike Fish from a nearby lake. Eleven months later, he complains of progressive weakness and fatigue. His CBC reveals a hemoglobin of 9.6 g/dL. What is the likely cause of his anemia?

a. Folate deficiency
b. Iron deficiency
c. Lead poisoning
d. Pyridoxine deficiency
e. Vitamin B12 deficiency

**Correct answer:** E

This man probably ingested larvae of Diphyllobothrium latum from raw freshwater fish. This fish tapeworm causes few abdominal symptoms, but can absorb vitamin B12 (choice E) to a great degree, causing a deficiency. This deficiency causes a macrocytic anemia.

Folate deficiency (choice A) can also cause a macrocytic anemia. Iron deficiency (choice B) and pyridoxine deficiency (choice D) cause a microcytic anemia.

Pyridoxine deficiency can cause anemia. But, there are neurological symptoms secondary to poor nerve conduction, especially in the hands and feet and it would be unlikely to be caused by the events in the vignette.
A 35 year-old male presents with fever and abdominal swelling. He reports weight loss and night sweats over the past month. Physical exam reveals enlarged abdominal lymph nodes; a lymph node biopsy demonstrates a "starry sky" appearance.

Which virus is most likely associated with the condition described above?

A. Epstein Barr Virus
B. Hepatitis B Virus
C. Human Immunodeficiency Virus
D. Human Papilloma Virus
E. Human T-cell Lymphoma Virus-1

Correct answer: A
This patient classically presents with Burkitt's lymphoma. This neoplasm has been linked to infection with Epstein Barr Virus (EBV, (A)).

(B) HBV is associated with hepatocellular carcinoma.

(C) HIV is associated with Kaposi's sarcoma.

(D) HPV is linked to cancers of the cervix and vulva, as well as laryngeal papillomas.

(E) HTLV-1 is linked to adult T cell leukemia and lymphoma.
A 5-year-old African-American boy with a history of sickle cell anemia presents with several days of left lower leg pain. The patient’s temperature is 102.3°F. Physical examination reveals a swollen, red area overlying the left tibia that is warm and tender to palpation. Which of the following organisms is most likely responsible for these signs and symptoms?

a. Escherichia coli  
b. Neisseria gonorrhoeae  
c. Salmonella  
d. Staphylococcus aureus  
e. Streptococcus pneumoniae

**Correct answer:** C

The correct answer is C. The patient presents with signs and symptoms of acute osteomyelitis, including localized erythema, swelling, tenderness, and warmth. The high-grade temperature is suggestive of the infectious nature of the disease. Although Staphylococcus aureus (Choice D) is the most common cause of osteomyelitis in the general population, Salmonella or other gram-negative organisms is the most common cause of osteomyelitis among patients with sickle cell disease. E. coli (Choice A), although a gram-negative bacteria, is not as common as Salmonella. Neisseria gonorrhoeae (Choice B) should be suspected in sexually active patients presenting with osteomyelitis and often manifests as septic arthritis. Streptococcus pneumoniae is a common cause of osteomyelitis in children younger than 24 months of age.
In clinic you see a 19-year-old college student who got back 10 days ago from a mission trip to Central America over Spring Break. Because he was only going to be gone for one week, he declined to take any malaria prophylaxis. For the last 4 days he has had fever, myalgia, chills, and nausea. Based on his history and clinical presentation, you diagnose him with malaria. Which stage of the protozoa infects red blood cells?

- a. hypnozoite
- b. gametocyte
- c. merozoite
- d. sporozoite
- e. trophozoite

**Correct answer:** C

The merozoite form (C) infects red blood cells.

Malaria is a vector-borne infectious disease caused by the protozoa Plasmodium, which has a sexual life cycle in the gastrointestinal tract of female Anopheles mosquitoes and an asexual life cycle in humans and primates. The female Anopheles mosquito injects sporozoites (D) into the blood stream. These invade the liver, where they develop in hepatocytes into schizonts full of merozoites. These are released to infect red blood cells, where they develop into ring forms and then into trophozoites (E). The trophozoites feed before returning to the schizont form. Some become merozoites while others become gametocytes (B), which when picked up by another feeding mosquito continues the chain of transmission. Plasmodium vivax and ovale may develop hypnozoites (A) in the liver, dormant forms that require primaquine to be eradicated. Plasmodium falciparum causes the most severe disease.
A patient has fever, malaise, rhinorrhea, and conjunctivitis for several days and thinks he has come down with the common cold. A few days later, he develops small, punctate, bluish-white dots on a red base on his tongue and this is followed by a maculopapular rash starting on face and spreading to torso and limbs. Which of the following is the most likely diagnosis?

a. Infection with a highly contagious, single-stranded RNA paramyxovirus
b. Infection with a tick-borne disease caused by Rickettsia rickettsii
c. Infection with a togavirus with an inner icosahedral capsid of RNA with an outer lipid envelope
d. Infection with a double-stranded DNA variola virus
e. Infection with Staphylococcus aureus and secondary release of toxins

**correct answer:** A
This patient has measles, which is an acute, infectious, and highly contagious viral infection caused by morbillivirus, a paramyxovirus, contacting the nasopharynx and bronchi and spreading to regional lymph nodes and systemically.

A tick-borne disease caused by Rickettsia rickettsii is Rocky Mountain Spotted Fever. The characteristic rash in this disease starts 3-4 days after onset of symptoms as pink flat macules on wrists and ankles. The rash then spreads to proximal arms, legs, torso, and finally to palms and soles before becoming hemorrhagic, petechial, and nonblanching.

An infection with a togavirus includes rubella or German measles. The characteristic rash in this infection also starts on the face and moves down the to the torso, but you would not see Koplik’s spots in this infection. In addition, rubella is usually subclinical and when it does present, it is often a milder infection than measles or rubeola.

Smallpox is an infection with a double-stranded DNA variola virus. The rash seen in this infection starts on the face, in the mouth, and on arms before spreading to the rest of the body. It is a macular rash that progresses to vesicles and pustular lesions in 1-2 weeks with scabbing. The only risk of smallpox is from bioterrorism, so this would be a much less likely cause of disease.

Infection caused by the release of toxins by Staphylococcus aureus is toxic shock syndrome. This infection results in acute onset of fever, vomiting, diarrhea, headache, sore throat, strawberry tongue, and erythematous mucous membranes (conjunctiva, oropharynx, vagina). The rash commonly seen is a scarlatina skin rash starting 5-7 days after symptom onset followed by desquamation occurring 7-10 days after onset on palms and soles. This is infection often has a rapid progression to multisystem organ dysfunction.
A 4-year-old girl came home from day care and developed fever, abdominal pain, and diarrhea containing flecks of bright-red blood and pus. The girl did not ingest any food at day care. Which is the most likely causative organism?

a. Enterotoxigenic Escherichia coli
b. Campylobacter jejuni
c. Shigella dysenteriae
d. Vibrio cholera
e. Yersinia enterocolitica

_correct answer: E_

Yersinia enterocolitica (choice E) occurs most often in young children, especially at day-care centers. Common symptoms include fever, abdominal pain, and diarrhea, which is often bloody. Symptoms typically develop 4 to 7 days after exposure and may last 1 to 3 weeks or longer.

Shigella dysenteriae (choice C) is a non-motile gram-negative rod. Humans are the only hosts for Shigella, and it usually causes bloody diarrhea.

Enterotoxigenic Escherichia coli (choice A) and Vibrio cholera (choice D) are both gram-negative rods that cause watery diarrhea.

Campylobacter jejuni (choice B) causes bloody diarrhea, but is usually due to ingestion of contaminated foods or water, with animals as the main reservoir. There is no given history of possibly contaminated food ingestion.
A 5-year-old child who has recently arrived from Sri Lanka is brought into the emergency room by her parents. They report that she has had a fever, headache, and a sore throat for the last two days. Physical examination reveals swollen lymph nodes in the neck, and a blood test indicates increased levels of serum amylase. The virus which is likely causing her infection is characterized by which of the following?

a. Double-stranded DNA genome  
b. Double-stranded RNA genome  
c. Negative-stranded RNA genome  
d. Positive-stranded RNA genome  
e. Single-stranded DNA genome

**Correct answer**: C

This child has a classical presentation of mumps. While vaccination for mumps is standard in industrialized nations, many children in developing nations do not receive the mumps vaccine. Mumps is caused by a paramyxovirus, which is a negative-stranded RNA virus. The serum amylase level is elevated due to parotitis - inflammation of the parotid gland. The other salivary glands (submandibular and sublingual) can be affected as well.
A 34 year old HIV+ male with a CD4 count of 85 develops confusion and a severe headache. He is found to be seizing by his roommate who brings him into the ER. At the hospital a contrast CT shows the ring enhancing lesions seen above. What was the most likely cause of this patient's current symptoms?

- a. Cat feces contamination
- b. Drinking unfiltered water
- c. Ingestion of cheese
- d. Ingestion of cured meat
- e. Reactivated infection

**Correct answer:** E

The patient is suffering from a Toxoplasma gondii infection. Given that his CD4 count <100 he is at risk for this infection. The most common source of infection in an immunocompromised host is reactivated infection (E). The past infection is never cleared from the system and when the host reaches a certain state of immune deficiency, reinfection is possible.

In developed countries primary infection is usually through ingestion of the oocytes in cat feces (from washing litter boxes, option A), or in cured/undercooked meat (option D). In developing countries, primary infection can happen from unfiltered water (option B).

Milk products (option C) are not a known source of toxoplasma infection.
A 35-year-old woman complains of migratory muscle and joint pain. Last month, she went hiking in the Adirondacks and noticed a single erythematous rash with a clear center a few days after returning. A few weeks later, additional rashes appeared and the patient reported onset of right knee pain. On physical exam, patient is notable for left sided facial drooping and left ocular ptosis. Blood serum analysis is positive for anti-Borrelia burgdorferi antibodies. What other pathogen is also carried by the tick that spreads *Borrelia burgdorferi*?

- a. Babesia microti
- b. Balantidium coli
- c. Leishmania chagasi
- d. Plasmodium falciparum
- e. Trypanosoma brucei

**Correct answer:** A

The *Ixodes scapularis* tick is the vector for *Borrelia burgdorferi* (Lyme disease) and protozoan *Babesia microti* (choice A). Babesiosis is similar to malaria in that the protozoan invades erythrocytes, causing fever and hemolysis. However, Babesia does not affect hepatocytes. The protozoan is spread by tick bites in northeastern United States, the same area that is endemic for Lyme disease. Also note that *Ehrlichia chafeensis*, a human pathogen, is also transmitted by the tick genus *Ixodes*; specifically species is *Ixodes persulcatus*.

*Balantidium coli* (choice B) is a protozoan spread by pig feces contamination. It can cause diarrhea.

*Leishmania chagasi* (choice C) is transmitted by the sandfly, causing visceral leishmaniasis (Kala-azar). The protozoan causes abdominal discomfort and distension, low grade fevers, anorexia, and weight loss.

*Plasmodium falciparum* (choice D) is responsible for malaria, which results in fever and hemolysis. It is spread by mosquitoes.

*Trypanosoma brucei* (choice E) is responsible for African sleeping sickness. It is transmitted by the tsetse fly.
During bacteriophage T7 infection of a single E. coli cell, 1000 bacteriophage particles are released. One of these particles contains a fragment of the E. coli chromosome rather than the T7 chromosome. Will this phage be able to inject its DNA into E. Coli cell, and if so, how many T7 particles will be produced by this cell?

Able to inject DNA # of T7 particles produced

a. Yes 1000
b. Yes 100
c. Yes 1
d. Yes 0
e. No 0

Correct answer: B
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Able to inject DNA #of T7 particles produced

a. Yes 1000  
b. Yes 100  
c. Yes 1  
d. Yes 0  
e. No 0

Correct answer: D
You are working at a pediatrics clinic when a mother brings in her 5-year-old son. She says that he and his brother had been suffering from "a cold" for the past week. Last night, he began crying inconsolably, holding his left ear, and she took his temperature to be 101.2. On exam today, you notice that his hearing is worse in his left ear. His mother says that his hearing was tested 3 months ago at his school and it was normal. When you are finally able to look into his left ear, you see a bulging, fluid-filled, erythematous tympanic membrane that does not move on pneumatic pressure. The child looks otherwise well, and the rest of his vitals are within normal limits. What is the most appropriate treatment?

a. Intramuscular amoxicillin  
b. Needle aspiration of the fluid for culture and pressure relief  
c. Observation and NSAIDs for pain relief  
d. Oral amoxicillin  
e. Oral ampicillin

**correct answer:** D

Most otitis media is caused by viruses, and antibiotics have been shown to reduce the natural disease course by only one day. Therefore, for most children, observation with NSAIDs for pain relief is sufficient. However, in this child with a history of an upper respiratory infection immediately preceding the onset of acute otitis media, a bacterial cause is suspected. Furthermore, the additional symptoms of fever and hearing loss should raise concern about this case of acute otitis media.

The two most common bacterial organisms causing AOM are *S. pneumoniae* and *H. influenza*, both of which are covered by amoxicillin. Because many strains of *H. influenze* and *M. catarrhals* are becoming resistant to ampicillin, this drug is no longer used as a first-line to treat ear infections.

Needle aspiration is both traumatic and unnecessary in this case. There is also no additional benefit to intramuscular amoxicillin over oral amoxicillin.
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A 31 year old man returning from a trip overseas presents with weakness. His body temperature is 38.1°C. His WBC count is 3,200; his hematocrit is 22.4, and his platelet count is 123,000. His skin examination reveals an ulcer. Upon further questioning he recalls that he was bitten by an insect at the ulcer site. Which of the following is the best treatment for this patient condition?

- a. Sodium stibogluconate
- b. Chloroquine
- c. Doxycycline
- d. Suramin
- e. Mebendazole

**correct answer:** A

This patient most likely has leishmaniasis, and the lesion described is the classic oriental sore which is formed at the skin area bitten by the sandfly, the transmission agent of leishmania. Patients usually present with pancytopenia, spiking fevers, and hepatosplenomegaly.

B is incorrect. Chloroquine is used in the treatment of malaria and amebiasis.

C is incorrect. Doxycycline is useful in the treatment of Lyme disease, malaria, anthrax, Chlamydia.

D is incorrect. Suramin is used to treat the blood-borne infection from trypanosomiasis, also known as African sleeping sickness.
Three days after recovering from the “flu”, a 35 year-old woman experiences numbness and tingling of the lower limbs. Over the following day, the numbness becomes accompanied by weakness and spreads to the upper extremities. Which of the following test results will confirm the presumptive diagnosis?

- a. Positive FTA-ABS test
- b. Positive Edrophonium challenge
- c. LP evidencing decreased glucose
- d. LP evidencing albumino-cytologic dissociation
- e. Positive culture for C. jejuni

correct answer: D

The patient evidences signs most consistent with Guillain-Barre syndrome (GBS), an autoimmune response activated by an infection. The hallmark finding in GBS is albumino-cytologic dissociation. The source of the increased protein count is the autoantibodies in the CSF.

Positive FTA-ABS (Fluorescent Treponemal Antibody ABSorbed) test indicates the presence of treponema pallidum, the causative agent of syphilis. Although syphilis does have manifest neurologically as tabes dorsalis -- degeneration of the dorsal columns resulting in paresthesias, locomotor ataxia, diminished reflexes, dementia -- these are not indicated by the question stem.

If the protein and cellular content of the CSF were increased proportionately (as opposed to albumino-cytologic dissociation), an infection rather than a post-infective syndrome would be more likely. Similarly, a lumbar puncture (LP) evidencing decreased glucose would indicate a bacterial infection as the bacteria would use the glucose normally present in the CSF for metabolism.

A positive edrophonium challenge (recovery of strength after edrophonium administration) would indicate myasthenia gravis (MG). While MG is also an autoimmune disease that strikes young women, it is not normally preceded by the infectious symptoms nor does it proceed from toe's to the head. Rather, the most classic presentation of MG is in weakness of the eyelids.

While, C.amylobacter jejuni is a typical agent that causes GBS, there are many reasons why a patient could have a positive culture for C.jejuni that do not involve GBS and there are many causes of GBS other than C. jejuni.
A 40 year old woman presents with new onset of seizures. She reports that she was having dinner with friends and suddenly lost consciousness. Her friends report that she fell to the ground and began moving her limbs in a rhythmic manner, lasting for about 30 seconds. She denies any history of neurologic problems. She is an immigrant from Ecuador and has been living in the US for the past ten years. Physical examination is unremarkable. A head CT is obtained. What is the most likely factor leading to her development of seizures?

- a. Consumption of undercooked pork
- b. Consumption of undercooked beef
- c. Consumption of fecally contaminated food or water
- d. Exposure to acid fast bacilli as a child
- e. Inheritance of gene encoding for an infective protein

**correct answer: C**

Neurocysticercosis is the most common cause of seizures in South America. Though it is acquired through infection from the pork tape worm Tenia solium (Option A), what actually leads to development of neurocysticercosis is consumption of eggs via the feces of a human infected by the parasite (through contaminated food or water, or poor hygiene). The mature worm lives in the jejunum of infected humans where it lays eggs. These eggs are passed with stool and are ingested by pigs, the intermediate host. Inside the pig, they hatch and migrate to different tissues including skeletal muscle. Undercooked pork meat infected with cysticerci is then eaten by humans, the definitive host, leading to the development of an adult worm in the jejunum. However, if humans consume the eggs (versus cysticerci in undercooked meat), it prevents them from completing their life cycle in the intermediate host and hatch entering the blood stream, never reaching the intestine. They can then disseminate to many areas of the body including skeletal muscle, eyes, heart, spinal chord and brain. They become encapsulated and are not able to mature into the adult worm. However, their presence causes an inflammatory reaction which can lead to many symptoms depending of the organ, including seizures.

Tenia saginata (Option B) is acquired from consumption of undercooked beef, and is not associated with cysticerci. Exposure to acid fast bacilli as a child (Option D) would suggest tuberculosis. While TB can affect the CNS, it would not produce the clinical and radiologic findings in this patient. Prion disease (Option E) would not cause the clinical picture presented on the case.
The strains of E.coli that cause urinary tract infections in patients uniquely express a structure that helps them bind to the uroepithelial cells. This structure is likely to be?

a. Sialic acid
b. Flagellum
c. Lactose
d. P pilus
e. Heat labile toxins

Correct answer: D
E.coli strains that cause UTI express a specific P pilus which binds to gal(1-4) on uroepithelial cells.
A 36-year-old man comes to the ER with a stiff neck and fever. He has had a sore throat and headache for the last 2 days and has vomited 3 times. On physical exam there are no focal neurological findings. On suspicion of meningitis, you order a lumbar puncture. While cleaning the skin over the lumbar vertebrae, you notice a raised, red insect bite on his lower back. If this patient turns out to have St. Louis Encephalitis, what kind of insect probably bit him?

a. Aedes mosquito
b. Anopheles mosquito
c. Culex mosquito
d. Dermacentor variabilis (American dog tick)
e. Ixodes tick

correct answer: C
C–St. Louis Encephalitis is due to a Flavivirus carried by Culex species of mosquito. (The same is also true of West Nile Virus and Japanese Encephalitis.)

A–Aedes mosquitoes carry the Flavivirus that causes Dengue Fever.

B–Anopheles mosquitoes transmit the Plasmodium species that cause malaria.

D–Rocky Mountain Spotted Fever follows the bite of a Dermacentor variabilis (American dog tick) carrying Rickettsia rickettsii.

E–Ixodes ticks carry Borrelia burgdorferi (Lyme Disease), Babesia microti (Babesiosis), and Erlichia ewingii (Ehrlichiosis).
A 47 year old woman presents with complaints of malaise and burning upon urination. She has a temperature of 39.6. Urinalysis is positive for nitrites and leukocyte esterase. A feature of the agent most likely responsible for this patient’s condition is:

a. A cell that is purple upon completion of the gram staining technique
b. A cell that is colorless after addition of ethanol during the gram stain procedure
c. An entity unable to live outside of a cell
d. A microorganism whose wall contains teichoic acid
e. A microorganism that requires an aerobic environment

**Correct answer:** B

This patient has a urinary tract infection (UTI) due to E. coli, the most common cause of UTIs. E. coli is a gram negative bacillus. From a stain point of view, the essential difference between gram positive and gram negative organisms is the fact that the former stains purple, whereas the latter stains pink. Addition of ethanol during step 3 (see table) washes away the crystal violet stain from the bacterial cells, as they have a thin of peptidoglycan cell wall.

There are four basic steps of the Gram stain, which include applying a primary stain (crystal violet) to a heat-fixed smear of a bacterial culture, followed by the addition of a mordant (Gram's iodine), rapid decolorization with alcohol or acetone, and counterstaining with safranin.

Answer A is incorrect; a purple cell at the end of gram staining is gram positive

Answer C is incorrect; it applies to viruses

Answer D is incorrect; teichoic acid is a polysaccharide found on the cell wall of gram positive bacteria

Answer E is incorrect; E. coli is a facultative anaerobe.
A young male was recently diagnosed with a sexually transmitted infection in which kidney-shaped, Gram-negative diplococci were found in cytoplasmic inclusions of neutrophils. The young male is allergic to penicillin. Administration of which of the following drug(s) is most appropriate?

a. Azithromycin  
b. Azithromycin & Doxycycline  
c. Ceftriaxone  
d. Ciprofloxacin  
e. Penicillin G with probenicid

**Correct answer:** B  
The patient has Neisseria gonorrhoeae. The current therapy of choice is ceftriaxone, but because of his allergy to penicillin, doxycycline (x7 days) is the best alternative treatment. However, the patient should also be treated with azithromycin for Chlamydia trachomatis because up to 50% of patients will be concurrently infected.  

Fluoroquinolones were frequently used for N. gonorrhoeae treatment until recently. In April 2007, the CDC discontinued the fluoroquinolone recommendation due to increased resistance among the population. See http://emedicine.medscape.com/article/218059-treatment
A sexually active 23-year-old female presents to clinic with complaints of red irritated eyes, dysuria, and diffuse joint pain. Pap smear is shown.

What organism is the most likely causative agent for her symptoms?

a. Chlamydia trachomatis
b. Gardnerella vaginalis
c. Plasmodium vivax
d. Rickettsia rickettsii
e. Trichomonas vaginalis

**correct answer:** A

This Pap smear demonstrates the characteristic cytoplasmic inclusions of the sexually transmitted obligate intracellular parasite Chlamydia trachomatis (Choice A). Her symptoms of conjunctivitis, urethritis, and arthritis are consistent with reactive arthritis. (Classic mnemonic: “Can’t see, can’t pee, can’t climb a tree.”)

Gardnerella vaginalis (Choice B) is the causative agent of bacterial vaginosis, a noninflammatory condition. It is diagnosed by its clinical characteristics: a thin white-yellow discharge; microscopy showing “clue cells,” epithelial cells covered with bacteria; positive “whiff test,” a fishy smell on KOH preparation; and a pH > 4.5.

Although Plasmodium vivax (Choice C) is a vector-transmitted intracellular parasite, it infects liver and erythrocyte, not cervical tissue. Signs and symptoms of infection include fever, headache, anemia, and splenomegaly.

Rickettsia rickettsii (Choice D) is a vector-transmitted intracellular parasite; though morphology is variable, it is characteristically Gram-negative, and would not typically be found in cervical tissue. It is the causative agent of Rocky Mountain Spotted Fever, with signs and symptoms of infection including fever, headache, and petechial rash with centripetal spread.

The sexually transmitted protozoan Trichomonas vaginalis (Choice E) is the causative agent of trichomoniasis, characterized by vaginitis, cervicitis, urethritis, and a green-yellow frothy discharge. It is classically diagnosed with a wet mount, not Pap smear, in which motile flagellated trophozoites are visualized.
A 22-year-old male presents to your clinic complaining of penile discharge and dysuria. The patient reports having multiple sexual partners but denies any history of sexually transmitted infections. Gram stain of his discharge reveals numerous neutrophils, but no bacteria. A feature of the organism causing your patient’s urethritis is:

- a. Antigenic variation
- b. Endoflagellum-mediated motility
- c. Host ATP-dependency
- d. IgA protease
- e. Thymidine kinase production

**Correct answer:** C

Host ATP-dependency (C) is a feature of Chlamydia trachomatis, an obligate intracellular bacterium (along with Rickettsia spp.). Extracellular elementary bodies (EBs) invade host cells and differentiate into reticulate bodies (RBs), which then replicate and differentiate back into the infectious EBs for release. This process takes place within inclusion bodies and is dependent upon host ATP.

Antigenic variation (A) and IgA protease (D) are both features of Neisseria gonorrhoeae, which would show Gram-negative diplococci on staining. N. gonorrhoeae not only variably express pili, but the pili genes undergo constant homologous recombination, thereby greatly contributing to immune system evasion.

Neisseria gonorrhoeae and Chlamydia trachomatis are the most common causes of urethritis in the US.

Endoflagellum-mediated motility (B) is a feature of Treponema pallidum (syphilis), which is not visible on Gram-stain, but would not likely produce penile discharge and dysuria. Instead, primary syphilis presents as a painless chancre. Secondary and tertiary syphilis would not likely cause penile discharge and dysuria.

Thymidine kinase production (E) is a feature of Herpesviridae Simplexvirus-2 (HSV-2), which is an important exploitation when treating with thymidine analogs such as acyclovir. HSV-2 would produce vesicular lesions.
A 29-year-old female complains of copious vaginal discharge and dysuria. On history, the patient confirms she has had previous sexually transmitted infections and uses an IUD for contraception. Gram stain of her discharge reveals numerous neutrophils and Gram-negative diplococci. You explain to your patient that after completing treatment she will not be protected against future infections from the organism causing this infection. A primary feature of this organism that facilitates evasion of adaptive immunity is:

a. A-B exotoxin production  
b. Beta-lactamase production  
c. IL-2 supression  
d. Pili gene variation  
e. Thymidine kinase mutation

**Correct answer:** D

Pili gene variation (D) is a primary contributor to immune system evasion of Neisseria gonorrhoeae. The constant homologous recombination of pili-associated genes allows the bacterium to alter its antigenic profile in a single generation. Additionally, N. gonorrhoeae can fail to express pili, thereby decreasing the number of cell-surface antigens. Opa gene expression, or failure to express, also contributes to antigenic variation. The implications of this include: failure to clear infection without treatment, difficulty in producing an effective immune response, difficulty in vaccine development. N. gonorrhoeae also produces an IgA protease, which would be another plausible answer to the question. IUD use is associated with an increased risk of gonorrhea.

A-B exotoxin (A) is not produced by Neisseria gonorrhoeae. Further, A-B exotoxins are not associated with immune system evasion, but rather host toxicity (i.e.: cholera, diptheria).

A beta-lactamase plasmid (B) can be associated with Neisseria gonorrhoeae. However it would result in drug resistance rather than adaptive immune system evasion.

Although IL-2 suppression (C) may diminish bacterial immunogenicity, Neisseria gonorrhoeae does not mediate this. In gonorrhea the immune system remains intact but pili antigenic variation facilitates evasion.

Thymidine kinase production (E) is an important feature of Herpesviridae Simplexvirus-2 (HSV-2), which is exploited when treating with thymidine analogs such as acyclovir. A mutation in the enzyme would cause drug resistance.
A sexually active 19-year old woman complains on increased frequency and a burning sensation upon urination. Pelvic examination shows inflammation of the exocervix and an exudate in the cervical os. Urinalysis shows numerous neutrophils but no bacteria. A cervical pap smear shows numerous lymphocytes and metaplastic squamous cells with vacuoles in the cytoplasm containing an inclusion. Which of the following pathogens is the causal agent?

- a. Candida albicans
- b. Chlamydia trachomatis
- c. Human Papillomavirus (HPV)
- d. Neisseria gonorrhoeae
- e. Trichomonas vaginalis

**Correct answer:** B

Answer Choice B - Chlamydia is correct

the patient has follicular cervicitis due to C.trachomatis. two distinct forms of the organism that develop in vacuoles withing metaplastic squamous cells are the elementary body (metabolically inert but infective) and the reticulate body (metabolically active but not infective) Binary fission of the reticulate bodies in the vacuoles of the infected cell results in the production of numerous elementary bodies. Urinalysis findings suggest acute urethral syndrome, which is also due to C.trachomatis. The treatment is a 1-g oral dose of Azithromycin

HPV is incorrect. HPV produces koilocytotic atypia of squamous cells, which is characterized by a halo surrounding a pyknotic(dense) nucleus. these cells are not described in the cervical pap smear

Gonorrhoeae also causes cervicitis and urethritis but is visible in gram stain gram negative diplococci. But they are not visible on Pap stains. Furthermore the pathogen does not produce inclusion bodies in teh metaplastic squamous cells.

Trichomonas are pear shaped organisms and have flagella, they are not described in the pap smear
A 31 year old man presents to the emergency department with increasing shortness of breath for the past three days, especially when climbing stairs or walking for a few minutes. He reports having fevers, chills, night sweats, and a non-productive cough. A chest radiograph demonstrates bilateral interstitial infiltrates. Laboratory studies reveal that the patient is HIV positive and has a CD4 count of 150. Additionally, the patient’s oxygen saturation is decreased to 92% on 3L of O2. What is the most likely causative organism of this acute illness?

a. Cytomegalovirus
b. Pneumococcus
c. Pneumocystis jiroveci
d. Tuberculosis
e. Histoplasmosis

correct answer: C

Patients with CD4 counts below 200 in the setting of HIV infection are considered to have AIDS despite viral titers. Immuno-compromised patients have increased susceptibility to opportunistic infections which includes several of the possible answer choices. However, only Pneumocystis carinii (choice C) fits the clinical picture. PCP is usually found in HIV patients with CD4 counts < 200 and the disease is characterized by constitutional symptoms, fever, night sweats, dyspnea on exertion, nonproductive cough, interstitial pattern on CXR, decreased PaO2, increased A-a gradient, and increased serum LDH.

Cytomegalovirus (choice A) is an opportunistic infection that has clinical manifestations of retinitis, esophagitis, colitis, hepatitis, and neuropathies.

Pneumococcus (choice B) is still the most common cause of pneumonia in immuno-compromised adults; however, it is characteristically a lobar pneumonia with discrete areas of consolidation on CXR as opposed to interstitial as described in this patient.

Tuberculosis (choice D) is unlikely in the setting of a non-productive cough and interstitial patterned infiltrates on CXR.

Histoplasmosis (choice E) has a similar clinical presentation as tuberculosis and would be equally unlikely in the setting of a non-productive cough and interstitial patterned infiltrates on CXR.
A 9 year old girl presents with 2 days of sore throat and fevers of 102ºF. Physical exam reveals an erythematous pharynx with a white, creamy exudate covering the left tonsil. Palpation of the neck reveals an extremely tender left submandibular lymph node. Throat cultures were taken and reveal beta-hemolytic colonies on blood agar. Susceptibility analysis show growth is not inhibited by amoxicillin, and erythromycin. Growth is inhibited by bacitracin. Which of the following is the mostly likely causal organism?

a. Rhinovirus
b. Streptococcus pyogenes
c. Streptococcus agalactiae
d. Epstein-Barr virus
e. Candida albicans

**Correct answer:** B

There are multiple organisms that can cause exudative pharyngitis; however, the results of the throat culture determine the final diagnosis. More specifically, Streptococcus pyogenes (choice B) can be correctly identified on culture based upon its characteristics of beta-hemolysis and susceptibility to bacitracin.

Rhinovirus (choice A) and Epstein-Barr virus (choice D) cannot be cultured on blood agar.

Streptococcus agalactiae (choice C) is a beta-hemolytic strep, but it is bacitracin-resistant and therefore not an appropriate answer. A more typical presentation of S. agalactiae is meningitis in newborns.

Candida albicans (choice E) can cause oral thrush; however, it seldom causes pharyngitis in non-immunocompromised individuals. Additionally, it does not grow beta-hemolytic colonies on blood agar.
A 54 year old health care worker tests 11 mm on his PPD exam that he gets annually. Chest X-ray does not demonstrate upper lobe cavitary lesion. He does not complain of any weight loss, fatigue or night sweats. Physical exam is normal. He receives isoniazid therapy for 9 months but starts to complain of stinging and burning sensations in his legs and arms a few weeks into his treatment. Supplementation with which of the following would resolve his new symptoms?

a. B6
b. Folate
c. B12
d. Vitamin A
e. Vitamin D

**correct answer:** A

This patient has B6 deficiency associated with isoniazid therapy for his TB. This patient has tested positive on PPD (> 11 mm for a healthcare worker is positive; > 5 mm for immunocompromised is positive, > 15 mm for normal people is positive) and thus needs INH therapy for 9 months due to his negative chest x-ray (for the treatment of latent TB). If his chest x-ray were positive he would need quadruple therapy for TB. INH alone is most commonly used when the PPD is positive and the chest x-ray is normal (latent disease). Its most common side effect is a peripheral neuropathy due to B6 deficiency. INH is known to interfere with b6 metabolism and should always be given w/ b6. While b12 can also cause neuropathy- it is not associated with INH use. Deficiency of b12 can cause subacute combined degeneration. The other vitamin deficiencies are not typically associated with neuropathy and INH treatment.
A 29 year old man with a CD4 count of 190 presents with a dry cough, fever for 2 weeks and shortness of breath, worsened when climbing stairs. His laboratory workup revealed a lactate dehydrogenase (LDH) level of 540. His blood pressure is 125/86. His chest X-ray is unremarkable. Which of the following is the most likely cause of the patient’s problems?

a. Congestive heart failure  
b. Pneumocystis jiroveci  
c. Escherichia coli  
d. Septic shock  
e. Streptococcus pneumoniae

**correct answer:** B

B is the correct answer

Pneumocystis jiroveci pneumonia (PJP, formerly known as Pneumocystis carinii pneumonia, PCP) occurs in immune compromised patients (CD4 below 200), often as an opportunistic infection in the setting of AIDS. It typically manifests as a non-productive cough and dyspnea worse on exertion, due to fibrosis of the lung parenchyma. A specific laboratory finding in PJP is an elevated LDH, often above 400. In fact, in a patient with an LDH below 220, PCP is very unlikely.

A is incorrect because of the patient’s young age. Also the clinical picture is more suggestive of an infectious process.

C is incorrect; when the CD4 count is below 200, opportunistic infections appear, but E coli is not a common cause. Also it is not known to cause a very elevated LDH.

D is incorrect, because in septic shock the patient is often hypotensive, which is not a finding in this case. Although the LDH would be low in septic shock secondary multi-organ failure, it usually is much lower.

E is incorrect, because S. pneumonia causes a productive cough. Additionally, the fever from S. pneumoniae would be more acute (less than 7 days), and would not cause an increase in LDH.
Which of the following is the most common cause of pneumonia in HIV patients?

a. Streptococcus pneumoniae  
b. Mycobacterium tuberculosis  
c. Pneumocystis jiroveci  
d. Histoplasma capsulatum  
e. Staphylococcus aureus

**correct answer:** A

AIDS patients, similarly to immunocompetent hosts, are most likely to get community-acquired pneumonia, mainly caused by bacterial organisms such as Streptococcus pneumoniae (the most common cause) and Haemophilus influenzae. B, C, D, E cause pneumonia in AIDS patients, but are not the most common causes.
A 3-week-old female presents to your clinic with cough, wheezes and fever lasting 1 day. Her mother states she was born at full-term without complications. History is negative for previous illness or significant familial diseases. Chest radiograph reveals diffuse hyperexpansion, bilateral interstitial infiltrates and bilateral upper lobe atelectasis. A major feature of the pathogen causing this infection includes:

- Complement system disruption
- Frequent antigenic shifts
- Fused respiratory epithelial cells
- Profound viremia
- Secretion of IgA protease

**Correct answer:** C

Fused respiratory epithelial cells (C) is a feature of Respiratory Syncytial Virus (RSV) also known as Paramyxoviridae pneumovirus. Epithelial infection is mediated by protein G; protein F causes infected cells to fuse and form syncytia, hence the name RSV. The most common cause of pneumonia in young children is RSV, followed by parainfluenza virus. Although bronchial smooth muscle is still forming in this infant, thus precluding traditional asthmatic wheezing, RSV is still able to cause bronchiolitis. Hyperexpansion, hypercapnia and hypoxemia

Complement system disruption (A) is a feature of Streptococcus agalactiae, or "Group B Strep" (GBS). The sialic acid moiety on the capsule binds factor H, which in turn accelerates degradation of C3b. This ultimately renders the alternative pathway complement activation inert. Although GBS can cause perinatal pneumonia, mothers are typically tested for vaginal GBS colonization.

Frequent antigenic shifts (B) can arguably be attributed to common flu virus (Orthomyxoviridae influenzae). However, influenza is not a common cause of pneumonia in children. Further, RSV antigenic shift is rare, if occurring at all.

Profound viremia (D) is not a feature of RSV; it rarely occurs.

Secretion of IgA protease (E) is a feature of some pneumonia-causing bacteria, such as Neisseria gonorrhoeae and Haemophilus influenzae. Like GBS, mothers are tested for STIs, thus diminishing the probability of Neisseria gonorrhoeae. Over 50% of Haemophilus infections present at meningitis. Other important manifestations include epiglotitis, cellulitis and septicemia.
A 66 year-old man presents complaining of fever and thigh pain. 24 hours ago he noted intense pain followed by redness and swelling in a small area of the right lateral thigh. Since then, the affected area has progressively increased to the current state, which on exam includes almost all of the right thigh and buttock, the lateral aspect of the right leg, and extends to the abdomen and right flank, 5 cm above the iliac crest. Crepitus is present on palpation. Leukocyte count is 14,000/mm3. Blood cultures are pending. Which organism is the most common etiologic agent of this clinical scenario?

a. Group A Streptococcus
b. Moraxella catarrhalis
c. Mycoplasma pneumoniae
d. Staphylococcus aureus
e. Staphylococcus epidermidis

**Correct answer: A**

This patient has necrotizing fasciitis. It is a rapidly progressive bacterial infection that spreads along fascial planes. It may follow trauma, but this will not always be present in the history. Crepitus is often appreciated, indicating subcutaneous air. Treatment includes starting IV antibiotics immediately, early and aggressive surgical debridement, and tetanus prophylaxis. It is classically caused by group A Streptococcus, but other etiologic organisms include Clostridium perfringens and Bacteroides fragilis (it is also often polymicrobial). Although Staphylococci are skin flora, they are not typically implicated in necrotizing fasciitis. Mycoplasma pneumoniae and Moraxella catarrhalis are causes of community-acquired pneumonia and do not cause necrotizing fasciitis.
A 16-year-old boy presents to you complaining of throat pain and malaise for the last week. His exam is significant for multiple palpable anterior cervical lymph nodes and WITHOUT tonsillar exudates. Three days ago his mother gave him leftover amoxicillin from a previous bacterial infection, and he subsequently developed a morbilliform, papular rash over his face, neck and trunk. Which of the following is the most likely cause of his symptoms?

a. Coxsackie A virus
b. Epstein-Barr virus
c. Group A streptococcus
d. Group B streptococcus
e. Measles virus

**Correct answer: B**

The Epstein-Barr virus is an extremely common cause of exudative pharyngitis and malaise in the teenage population. Additionally, 90% of those infected with EBV who take amoxicillin will develop a measles-like "amoxicillin rash," making it the leading diagnosis after the history and physical. This same rash is also associated with ampicillin.
A 22-year-old college student presents to Student Health during his final exams complaining of a painful, vesicular lesion on his upper lip as seen in the accompanying image. He gives a history of having had similar lesions in the past in the same location. What type of organism is most likely responsible for the student’s symptoms?

- A single-stranded RNA virus
- A double-stranded RNA virus
- A double-stranded non-enveloped DNA virus
- A single-stranded non-enveloped DNA virus
- A double-stranded enveloped DNA virus

**Correct answer:** E

The lesion seen in the photo represents herpes labialis from infection with herpes simplex type I infection (HSV-I). HSV-I is part of the herpesviridae family and is an enveloped double-stranded DNA virus. Other members of the herpesviridae family include herpes simplex type II (HSV-II), Varicella-Zoster virus, Epstein-Barr virus, and cytomegalovirus. The virus remains latent in the trigeminal root ganglion and reactivation is provoked by stress or illness resulting characteristically in lesions in the same location as previous outbreaks.

A. Most RNA viruses are single-stranded including the families calicivirus (Norwalk), picornavirus (polio, enteroviruses, hepatitis A), flavivirus (yellow fever, hepatitis C), togavirus (rubella).

B. An example of a double-stranded RNA virus is rotavirus, the most common cause of acute viral gastroenteritis in infants and children.

C. Double stranded naked DNA virus include adenovirus. Adenovirus serotypes 11 & 21 are associated with hemorrhagic cystitis in child care. Adenovirus serotypes 4 & 7 used to be provided to the military recruits as vaccines. Increase incidence of adenovirus occurs with military recruits and day care centers.

D. An example of a single-stranded DNA virus is parvovirus B19. This agent causes erythema infectiosum (Fifth disease) in children and is linked to aplastic crises in sickle cell anemia patients. All other DNA viruses are double-stranded.
A 23 year old female presents to the ER with nausea, vomiting, photophobia, rash, and a fever of 102.0 F. Two days prior to admission, a diffuse maculopapular rash erupted on her wrists and progressively spread to include her trunk, back, chest, and abdomen. PE exam is notable for a petechial rash on the extremities and chest, as well as conjunctivitis present bilaterally. Upon further history, patient revealed that she had recently gone camping. What is the most likely diagnosis?

a. Lyme Disease
b. Rocky Mountain Spotted Fever
c. Meningococcemia
d. Erythema Multiforme
e. Ehrlichiosis

correct answer: B

This patient likely has RMSF. She is presenting with the classical signs of RMSF including nausea and vomiting, photophobia, and a centripetal rash. A centripetal rash is one that starts on the wrists and works its way towards the rest of the body. The petechial rash and conjunctivitis are also classic for RMSF; as is the history of camping. The treatment for RMSF is Doxycycline. When camping is mentioned think of RMSF or Lyme. Lyme disease is likely to present with a large bulls eye lesion called erythema chronicum migrans. The disease can progress to include a bilateral Bell’s Palsy and cardiac and joint manifestations may surface at a later point if treatment is delayed. Treatment is with Doxycycline; ampicillin or amoxicillin can be used as alternatives. Meningococcemia is possible and may also present with a petechial rash and fever- but the history of camping and the centripetal nature of the rash is more like RMSF. Always be wary of acute hemorrhage into the adrenal glands with Meningococcemia. Erythema Multiforme is filler here and is a target shaped lesion often seen with drug reactions and mycoplasma/HSV infections.
The stuffed animal in _The Velveteen Rabbit_ is condemned to be burned with the other nursery toys after its owner comes down with sore throat, a confluent erythematous sandpaper-like rash, and elevated temperature. What causes the rash in scarlet fever?

a. desquamation from exfoliative toxins A or B  
b. desquamation from TSST-1  
c. desquamation from streptococcal pyrogenic exotoxin (SPE)  
d. CD8+ T cells killing infected vascular endothelial cells  
e. delayed hypersensitivity reaction to SPE

**Correct answer:** E

Scarlet fever results from infection with Group A beta-hemolytic streptococci, namely Streptococcus pyogenes. Clinical presentation may include fever, sore throat, headache, nausea, vomiting, malaise, strawberry tongue, and this non-infectious, autoimmune rash, a delayed (type IV) hypersensitivity reaction to the streptococcal pyrogenic exotoxin (SPE).

A--Desquamation from exfoliative toxins A and B is called Staphylococcal Scalded Skin Syndrome. This condition, usually seen in infants, tends to be less serious than it looks!

B--Desquamation from Toxic Shock Syndrome Toxin 1 (TSST-1) is seen with some Staphylococcal infections; it may also result from SPE (C), but this is not what causes the rash of scarlet fever.

D--CD8+ T cells killing infected vascular endothelial cells causes the erythematous maculopapular rash of measles that begins at the hairline, spreads down the body, and may desquamate.
An 8-year-old child with cough and fever has a maculopapular rash spreading from her hairline down her face and the rest of her body. The rash is due to cytotoxic T cells killing infected epidermal skin cells and is not contagious. What microorganism is most likely causing her illness?

a. A double-stranded, enveloped DNA virus
b. A Gram-positive bacterium growing in clusters
c. A Gram-positive diplococcus
d. A single-stranded positive, non-enveloped RNA virus
e. A single-stranded negative, enveloped RNA virus

**Correct answer:** E

This patient demonstrates the classic rash of measles, which is due to E--A single-stranded negative, enveloped RNA Paramyxovirus. It is spread in respiratory droplets and causes transient immune suppression (weeks-months). Most measles deaths are due to bacterial superinfection; rarely there is encephalitis, deafness, or mental retardation. International efforts to strengthen mucosal defenses with Vitamin A have decreased measles incidence worldwide.

A--A double-stranded, enveloped DNA virus (like Varicella Zoster Virus, which causes chicken pox and shingles; or Human Papilloma Virus or Herpes Simplex Virus)

B--A Gram-positive bacterium growing in clusters (like Staphylococcus aureus, which causes impetigo and Scalded-Skin Syndrome)

C--A Gram-positive diplococcus (like Group A beta-hemolytic Streptococcus pyogenes, which causes rheumatic fever and scarlet fever)

D--A single-stranded positive, non-enveloped RNA virus such as the enteric Picorna Viruses (like Coxsackie Virus)
A 35 year-old woman had palpitations and anxiety. She also had double vision and decreased visual acuity, and was subsequently successfully treated with Iodine-131 and surgery on the soft tissues surrounding the eyes. She is now 54 years old, is asymptomatic, and takes replacement therapy. The mechanism of action of the hormone that is being replaced is which of the following?

a. The hormone is normally secreted by the anterior pituitary gland, its receptors on the thyroid gland have 7 transmembrane domains and, through various signaling cascades, acts to upregulate gene transcription.

b. The hormone is normally secreted by thyroid epithelial cells, and much of it binds to thyroid binding globulin in the plasma. Much of the hormone is deiodinated, and its target is in the nucleus of receptor cells and it acts to upregulate gene transcripti

c. The hormone is normally stored in colloid of the thyroid gland and is secreted by thyroid interstitial cells after deiodination by the thyroid epithelial cells; the hormone then acts on a tyrosine kinase receptor on target cells which, through a series of

d. The hormone is a tripeptide hormone normally stored in the colloid of the thyroid gland. Upon stimulation by TSH, the hormone is cleaved from thyroglobulin and secreted through the thyroid epithelial cells into the bloodstream. It acts on nuclear recept

e. The hormone is made in the hypothalamus and travels intracellularly to the posterior pituitary gland (neurohypophysis) where it is stored. It is released when plasma osmolality increases or blood pressure significantly decreases and it acts on the kidney

**correct answer:** B

The answer is B. The patient had Graves disease when she was 35. Graves disease is an autoimmune disease in which autoantibodies bind to the TSH receptors on thyroid epithelial cells and stimulate release of thyroxine, leading to a hyperthyroid state. Young women are often affected and symptoms may include anxiety, palpitations, diaphoresis, heat intolerance and patients often have an increased basal metabolic rate possibly leading to weight loss. Exophthalmos is an effect mostly occurring in Graves disease. Mucopolysaccharides are deposited in the extraocular muscles as well as in the soft tissues surrounding the eyes, which may lead to visual problems, and in severe cases may require corrective paraocular surgery.

The hormone being replaced here is thyroxine, T4, or thyroid hormone, and this is usually done with the drug levothyroxine. T4 is made in thyroid epithelial cells and stored as thyroglobulin in colloid. It is secreted upon stimulation from TSH. It binds thyroxine binding globulin in the periphery and much of it is deiodinated in the periphery to T3, the more abundant unbound form of the hormone. It acts intracellularly in the nucleus of target cells and binds directly to transcription factors, leading to upregulation of gene transcription.

Choice A is incorrect; thyroid stimulating hormone is secreted by the anterior pituitary and binds to a G protein coupled receptor on thyroid epithelial cells, leading to thyroid hormone release.

Choice C is incorrect; the hormone is not secreted by thyroid interstitial cells, rather it is secreted by epithelial cells. Also, its receptor is not a tyrosine kinase receptor, it is a nuclear receptor.

Choice D is incorrect; thyroxine is not a tripeptide hormone, it hydrophobic. TRH is a tripeptide hormone and is secreted from the hypothalamus and acts on receptors in the anterior pituitary.

Choice E is incorrect; this choice describes antidiuretic hormone.
A 51-year-old woman with cancer is being treated with a hematopoietic growth factor. Leukocyte differentials before and after treatment are shown above. The growth factor is most likely to be which of the following?

- a. Granulocyte colony-stimulating factor
- b. Transforming growth factor-beta
- c. Interleukin-6 (IL-6)
- d. Interleukin-8 (IL-8)
- e. Macrophage colony-stimulating factor

**Correct answer: A**

Granulocyte colony-stimulating factor (choice A) increases neutrophil, eosinophil and basophil counts: the patient’s white blood cell differential shows an increase in the percentage of neutrophils from 9% to 90% after the growth factor was given. The percentage of eosinophils did not change, but since the total number of cells increased, the absolute eosinophil amount also increased. The percentage of lymphocytes decreased.

The other growth factors listed would not increase production of neutrophils. Transforming growth factor-beta (choice B) is made by macrophages, endothelial cells, and T-cells and is involved in B-cell maturation.

IL-6 (choice C) is an acute phase reactant made by macrophages, and

IL-8 (choice D) is involved in neutrophil migration and chemotaxis.

Macrophage colony-stimulating factor (choice E) would increase the percentage of cells in the monocyte-macrophage line, but that information is not provided in the question.
A first-year medical student is new to the lab and wants to know if a well-studied gene is transcribed in the sample of cytoplasmic extracts she is given. Which of the following techniques should she use to quickly find out?

a. Column chromatography
b. Northern blot
c. Southern blot
d. Southwestern blot
e. Western blot

**Correct answer:** B

Northern blot employs a hybridization probe (of the gene of interest) to identify complimentary RNA sequences. Southern blot is used for hybridization to DNA sequences (which would not tell you anything about expression). Western blot is used for protein identification (not gene expression). Southwestern blot is used to determine proteins that bind to DNA. And column chromatography is used to fractionate extracts based on a number of factors (size, charge, pH, etc.), but won't identify individual genes.
A segment of DNA has the following structure, complete with restriction enzyme recognition sites: -[5kb]-|EcoRI|--[10kb]--|TaqI|---[15kb]---|EcoRI|-[5kb]-|BamHI|--[10kb]--

This segment is replicated using PCR and sequentially subjected to different restriction endonucleases. The resulting mixture is separated using gel electrophoresis. If bands are found at only 5kb, 10kb, and 25kb lengths, which restriction enzymes were utilized?

a. BamHI only  
b. EcoRI and BamHI  
c. EcoRI and TaqI  
d. EcoRI only  
e. EcoRI, BamHI, and TaqI

Correct answer: B

First, one must understand that splicing a DNA segment at a restriction enzyme recognition site results in two DNA segments. Apply this principle to multiple sites with multiple enzymes and you end up with a slew of different DNA segment lengths. The key is that the size of the ending segments depends upon the combination of restriction enzymes that are applied. EcoRI alone results in bands of 5kb, 15kb, and 25kb. BamHI alone results in bands of 10kb and 35kb. EcoRI and BamHI leads to bands of 5kb, 10kb, and 25kb (correct answer - don’t forget the double 5kb band). EcoRI and TaqI lead to bands of 5kb, 10kb, and 15kb. When all three enzymes are employed, the result is bands at 5kb, 10kb, and 15kb.
The locations of three different nucleotide substitution mutations near the human beta-globin gene are shown by the vertical arrows. Each of these mutations can lead to beta-thalassemia major. These three nucleotides of the wild-type sequence are most likely to be required for which of the following?

- Binding eIF4F
- Binding eIF4G
- Binding histone acetylase
- Binding histone deacetylase
- Binding positive-acting transcription factors

**Correct answer: F**

To answer this question, you need to know that beta-thalassemia major is caused by under-production of the beta globin protein, the product of the beta-globin gene. Since all three of the nucleotides in this question are upstream of the transcription start site, they are not part of the gene transcribed, and therefore are not required for binding elongation factors, such as eIF4F (choice A) or eIF4G (choice B), which are involved in translation of proteins, not the transcription.

Similarly, histone acetylase and deacetylase (choices C and D) are involved in chromatin packing of the gene as a whole, not a particular region, as in this example.

A loss of function mutation in positive-acting transcription factors (choice F) is the only choice that would result in under-production of the entire protein product by affecting the promotor region.
A 30 year old man comes to emergency room complaining of right upper quadrant pain. He had just returned from a two week trip to Italy. One day after taking a tour of the Italian countryside, he began having diarrhea and abdominal pain that subsided within 24 hours. On physical exam, he is ill appearing and short of breath. He has a moderate amount of jaundice. Upon further questioning, he admits to eating mushrooms while on his tour of the Italian countryside. A urinalysis is performed and he is found to have alpha-amanitin, an inhibitor of RNA polymerase II. The synthesis of which of the following would most likely be affected?

- a. cDNA
- b. DNA
- c. mRNA
- d. rRNA
- e. tRNA

correct answer: C

Alpha-amanitin is found in death cap mushrooms. These types of mushrooms are native to Europe and are widespread. Early symptoms of intoxication are diarrhea and cramping which subside within the first day. This can give a false sense of remission. Intoxication eventually leads to renal failure, respiratory failure, and eventually death within the first week after ingestion. RNA polymerase II is responsible for the synthesis of mRNA (choice C). RNA polymerase I makes rRNA (choice D) and RNA polymerase III makes tRNA (choice E).

RNA polymerases do not make DNA (choice B).

cDNA (choice A) is not naturally occurring in the body. It is made in the laboratory and is a copy of all mRNA encoding regions present in cells. It is tissue specific and expresses genes found only in the tissue from which it is derived.
A patient with a rare hereditary tumor syndrome is found to have a mutation in a gene on chromosome 16. When researchers clone the gene, they find that the mutant gene generates a protein that is 428 amino acid residues long. The wild type of the gene normally produces a protein 683 amino acid residues long. The researchers note that the mRNA generated from the mutant gene is significantly longer than the wild type. In which part of the gene did the mutation likely take place?

a. Enhancer  
b. Operon  
c. Splice Site  
d. Promoter  
e. Start codon

**Correct answer:** C

This patient has a splice site mutation. The splice site is located within the intron. The removal of the splice site causes the mutant gene to retain an intron and results in the production of an aberrant protein. In this case, the added intron contains a stop codon and causes the translation of the protein to terminate at codon 428. This explains why the mRNA is longer than normal but the protein is shorter.

A mutation in the enhancer or the promoter may cause weaker expression of the mutant protein but would not affect its length.

A mutation in one of the exons may create a stop codon which causes translation to terminate early, but would not create a longer mRNA.

A mutation in the start codon would cause the mutant protein not to be expressed at all.
A 18-year-old white female has a history of recurrent pneumonia since birth and her brother also has this disease. She regularly undergoes chest physiotherapy, takes pancreatic enzymes, bronchodilators, and mucormyst (n-acetylcysteine). These observations suggest that the multi-organ disease described above is as a result of which of the following genetic principles?

- a. Anticipation
- b. Imprinting
- c. Incomplete Penetrance
- d. Locus Heterogenity
- e. Pleiotropy

**Correct answer:** E

The answer is E. Pleiotropy is defined by a single disease causing mutation that affects multiple organ systems, it is common among genetic diseases and Marfan syndrome and Cystic Fibrosis (as described in the vignette above) are just a few examples.

(A) Anticipation is when individuals from generation to generation get the disease at an earlier age and the severity worsens. These are commonly associated with trinucleotide repeat diseases such as Huntington disease (AD), Fragile X (XD), and Myotonic Dystrophy (AD).

(B) Imprinting is when genes are only "transcriptionally" active when it is specifically transmitted by either the mother or father; examples include Prader-Willi and Angelman Syndromes. Can be remembered as: Mother is an Angel.

(C) Incomplete Penetrance is defined when there is no phenotypic expression and thus no pathology. Note that this is different from variable expression because in this every diseased person will have some level of the disease (e.g. hemochromatosis, Neurofibromatosis).

(D) Locus Heterogenity is simply when mutations at different loci can produce the same phenotype (e.g. albinism, osteogenesis imperfecta)
A 64 year-old man presents with 2 days of left knee pain. He is afebrile and has no signs of systemic infection. He denies trauma to the knee and states that the knee looks swollen and red and feels hot and extremely painful. A knee aspirate is done and polymorphonuclear cells within the joint space are found to contain crystals that are negatively birefringent under polarized light. If the patient is allergic to indomethacin, what is the mechanism of action for the medication you prescribe

a. Inhibition of xanthine oxidase and decrease in production of uric acid
b. Inhibition of tubulin polymerization into microtubules by binding to tubulin
c. Inhibition of prostaglandin synthesis by irreversibly binding COX-1
d. Stabilization of microtubules to prevent mitosis in rapidly-dividing cells
e. Inhibition of production of the bacterial cell wall peptidoglycan cross links resulting in a bacteriocidal effect

**Correct answer:** B

**Answer:** B. The patient is having an acute attack of gout. Gout is a crystal disease that is diagnosed when PMNs aspirated from such a knee show intracellular crystals which are negatively birefringent under polarized light. For acute attacks, NSAIDs may be used for pain and colchicine is commonly used as well. The mechanism of action of colchicine is by binding to and inhibiting tubulin assembly in cells. This inhibits migration of neutrophils into the joint space and decreases the inflammatory response by decreasing mitotic divisions.

Choice A is incorrect; this mechanism describes allopurinol which may be used for long term treatment in gout patients but should never be started or stopped during an acute attack because any change in uric acid metabolism may exacerbate the attack.

Choice C is incorrect; this mechanism describes aspirin, not colchicine

Choice D is incorrect; this mechanism describes docetaxel which is used mainly in the treatment of breast and ovarian cancers

Choice E is incorrect; this mechanism describes penicillin which is not indicated in this synovial crystal disease. Antibiotics are indicated in a septic joint.
During the proliferative phase, plasma estrogen levels rise and estrogen receptors in the endometrium increase in number. What is the most likely mechanism by which the number of estrogen receptors increase?

- a. Increased hepatic lysosomal enzyme synthesis
- b. Increased transcription of pregnenolone
- c. Increased phosphorylation of amino acid residues
- d. Mitosis
- e. Proteolytic cleavage

**Correct answer:** D

Like all steroid hormones, estrogen works at the level of DNA, and so mitosis (choice D), which increases DNA, necessarily increases the number of available estrogen receptors. The other type of estrogen receptor is GPR30, an integral membrane protein.

Increased hepatic enzyme synthesis (choice A) increases protein, but estrogen receptors are made of DNA. Increased transcription of pregnenolone will increase estrogen precursor levels without altering estrogen receptor availability (choice B), but estrogen receptors are made of DNA. Increase phosphorylation of amino acid residues (choice C) is a modification of protein, but estrogen receptors are made of DNA. Proteolytic cleavage (choice E) is not the mechanism by which estrogen activates DNA for transcription.
A 60 year old man with a history of myocardial infarction is receiving an echocardiogram to assess the ejection fraction of his left ventricle. It is discovered that the patient has a dilated left atrium with a visible wall thrombus. He is then started on anticoagulation therapy. Which of the following organs will receive a white (pale) infarct if the left atrial thrombus embolizes?

a. Kidney  
b. Large intestine  
c. Liver  
d. Lung  
e. Small intestine

**correct answer:** A

An infarct describes an area of tissue undergoing coagulative necrosis resulting from decreased blood supply. Embolization of a thrombus can occlude arterial vessels, resulting in infarcted tissue in various organs. Infarcts can be classified into either red (hemorrhagic) infarcts or white (pale) infarcts based upon their gross appearance on pathology. Red infarcts usually occur in loose tissues with collateral circulation or following reperfusion of the infarcted tissue. Pale infarcts usually occur in solid tissues with a single blood supply. Of the listed organs, only the kidney (choice A) has a single blood supply without collateral circulation. Liver (choice B), Lung (choice C), Small Intestine (choice D), and Large Intestine (choice E) all have more than one blood supply. An infarct in these tissues would tend to be grossly red on pathological examination.
A 6yr old child presents with flu-like illness and palpable rash on buttocks and thighs, he also complains of abdominal pain and multiple joint pains. Urine examination revealed haematuria and casts. What is the likely pathology of this disease?

- a. Type II Hypersensitivity IgM mediated
- b. Type II Hypersensitivity IgG mediated
- c. Type III Hypersensitivity IgG mediated
- d. Type III Hypersensitivity IgA mediated
- e. Type III hypersensitivity IgM mediated

**Correct answer:** D

Palpable Rash is a characteristic finding of small vessel vasculitis, and the common vasculitis in children is Henoch Schönlein purpura which has a palpable purpura due to type III hypersensitivity producing IgA immunocomplexes.
A 28 yrs old man is brought to ER because of fatigue and fever of acute onset. He has a history of I/V drug use. Echo suggests endocarditis of the tricuspid valve. He has no past history of valvular heart disease. Blood cultures are likely to grow

a.  S. epidemitis  
b.  S.aureus  
c.  S.pyogenes  
d.  T.pallidum  
e.  S.mutans

**correct answer:** E
Viridans group streptococci are alpha hemolytic, most common cause of bacterial endocarditis. They are resistant to optochin.
The most common cause of hydrops fetalis is
a. Trisomy 21
b. Alpha thalassemias
c. B19
d. Twin to twin transfusion
e. High output failure

**correct answer:** E

The most common cause of hydrops fetalis is cardiovascular malformations leading to high cardiac output failure. This is a highly tested on the boards because a lot of students always think about alpha thalassemias.
A 35 year old professional soccer player suddenly collapses and dies during a soccer match. What pathological finding will most likely be found on autopsy?

- a. Coarctation of aorta
- b. Dilated cardiomyopathy
- c. Hypertrophic cardiomyopathy
- d. Restrictive cardiomyopathy
- e. Ventricular septal defect

**Correct answer:** C

The correct answer is C: Hypertrophic cardiomyopathy. 50% of the cases of Hypertrophic cardiomyopathy are familial and are inherited as an AD trait. It is a common cause of sudden death in young athletes. The ventricles and the septum become hypertrophied to the point where the ventricle becomes banana-shaped.
A 32-year-old man presents to the emergency room with headache, palpitations, sweating, and anxiety for the past 30 minutes. His blood pressure is 220/110 mm Hg. Upon questioning the patient reports that he’s experienced similar episodes intermittently over the last year, with complete resolution of symptoms between episodes. Further testing in this patient will most likely show which of the following?

a. Decreased aldosterone  
b. Decreased blood glucose  
c. Elevated aldosterone  
d. Elevated plasma free metanephrine  
e. Elevated renin

**Correct answer:** D

This patient likely has a pheochromocytoma. The classical presentation is paroxysmal hypertension associated with the triad of headaches, sweating, and palpitation. The best test for confirming pheochromocytoma is a finding of elevated plasma free metanephrine. The most practical and best confirmatory test for Pheochromocytoma is Urine VMA assessment.
A 40 year old woman has experienced lethargy weakness and constipation for past 6 months. Physical examination is normal except for tenderness of left 3rd proximal finger and slightly irregular heart beat. An ECG shows prolonged QT (Corrected) interval. Lab shows calcium 11.6mg/dl, phosphorus 2.8mg/dl and alkaline phosphatase 202U/L. Radionuclied scan shows 1cm area of increased uptake in the right lateral neck. The most likely gene mutation is

- a. MEN1
- b. GNAS1
- c. RET
- d. p53
- e. NF1

**Correct answer:** A

This patient has parathyroid neoplasm most likely adenoma with hypercalcemia complicated by ostietis fibrosa cystica of her finger. MEN 1 is the most common mutation in the parathyroid tuours. MEN 1 is a tumour suppressor gene, the loss of which not only occurs in sporadic parathyroid tumours but also in multile endocrine neoplasia 1. GNAS1 is mutated in pituitary adenomas. RET in medullary carcinoma thyroid.
A 25-year-old man presents to the emergency department with fever, jaundice, and pain localized to the right upper quadrant. Subsequent endoscopic retrograde cholangiopancreatography (ERCP) findings show alternating strictures and dilation of the bile ducts with a characteristic "beading" appearance. Both intrahepatic and extrahepatic bile ducts are involved. What is the most likely diagnosis?

a. Crohn's disease  
b. Polymyalgia rheumatica  
c. Primary sclerosing cholangitis  
d. Sarcoidosis  
e. Systemic lupus erythematosus

correct answer: C  
Primary sclerosing cholangitis (PSC) is a chronic idiopathic disorder involving inflammation and fibrosis of both intra- and extrahepatic bile ducts. Alternating dilation and stricture of bile ducts results in characteristic "beading" appearance on ERCP. Up to 70% of patients with PSC suffer from ulcerative colitis.
A 19-year-old college student has chronic bouts of abdominal pain and diarrhea. On many occasions she has seen blood in her stool. Her sister has the same condition. Which of the following is a feature of the most likely diagnosis in this case?

- a. Cobblestone mucosa
- b. Crypt abscesses and ulcers
- c. Noncaseating granulomas
- d. Skip lesions
- e. Transmural involvement of the small bowel

**Correct answer:** B

This patient has symptoms of inflammatory bowel disease. The presence of bloody stools and a positive family history should make you lean toward ulcerative colitis over Crohn's.

Crohn's disease is marked by transmural involvement, skip lesions, noncaseating granulomas, and cobblestone mucosa.

UC is marked by mucosal inflammation, continuous involvement usually in the descending colon and rectum, crypt abscesses and ulcers and a friable mucosa with pseudopolyp formation.
A 45 year old male comes to the emergency department complaining of severe abdominal pain and non-bloody diarrhea. On examination he has fever. He was recently in the hospital for 7 days for treatment of pancreatitis, and was given antibiotics during his stay. He has not traveled recently. What is the most likely cause of this man’s diarrhea?

- a. Crohn’s disease  
- b. Enterotoxic E. coli  
- c. Irritable bowel syndrome  
- d. Pseudomembranous colitis  
- e. Ulcerative colitis

correct answer: D

This man most likely has C. difficile colitis following his hospital stay and antibiotic treatment. Pseudomembranous colitis is characterized by diarrhea, abdominal pain (which can be severe), and fever.

(B) This man has not traveled recently, which makes this diagnosis less likely.  
(C) The clinical history is not consistent with IBS.  
(A,E) This man does not have systemic symptoms that would suggest inflammatory bowel disease. The proximity to his hospital stay and antibiotic treatment makes C. difficile colitis more likely.
A 58-year-old man with a history of epigastric tenderness and diarrhea is found to have a peptic ulcer and hypertrophic gastric fold on upper GI series. Using somatostatin receptor scintigraphy, a hormone-secreting tumor is identified in the pancreas. Given the most likely diagnosis, which of the following laboratory results would be expected?

- Decreased d-xylose test
- Fasting serum gastrin >1000pg/mL
- Increased 5-HIAA in urine
- Positive urea breath test
- Serum antiparietal cell antibodies

**Correct answer:** B

The correct answer is fasting serum gastrin >1000pg/mL. This man has Zollinger-Ellison syndrome, which is a pathological condition caused by gastric acid hypersecretion secondary to a gastrinoma. In these patients the fasting serum gastrin level is abnormally elevated. Further investigation would reveal an increase in gastrin during the secretin-stimulation test. These patients are at risk for gastric bleeding and duodenal perforation.

The other choices are laboratory findings in other gastrointestinal diseases. Increased 5-HIAA in urine is seen in patients with Carcinoid tumor. Patients with gastritis secondary to H. pylori infection often have a positive urea breath test. Serum antiparietal cell antibodies are seen in patients with pernicious anemia. Finally, decreased d-xylose test can be found in patients with Celiac Sprue, a disease characterized by resolution of intestinal lesions and malabsorption with avoidance of dietary gluten.
A 19 year-old woman presents to the emergency room with intense right lower quadrant abdominal pain of 2 hours duration. On physical examination, the patient has a temperature of 38.5°C (101.3°F) and blood tests reveal a WBC count of 13,000/mm3. Which symptom is the patient also likely to be suffering from?

- Abdominal distention
- Hunger
- Nausea and vomiting
- Profuse watery diarrhea
- Vaginal bleeding

**Correct answer:** C

The correct answer is C. New onset of right lower quadrant abdominal pain in a young patient with a fever and an elevated WBC count is suggestive of appendicitis. Patients with appendicitis typically present with periumbilical or right lower quadrant abdominal pain. Fifty to sixty percent will also present with nausea and vomiting.

A is not correct. Distention is rare in a patient with appendicitis unless diffuse peritonitis has developed.

B is not correct. Patients with appendicitis typically present with anorexia. Hunger in a patient with appendicitis should make one reconsider the diagnosis.

D is not correct. While diarrhea can occur in a patient with appendicitis, profuse watery diarrhea is more suggestive of acute gastroenteritis than appendicitis.

E is not correct. Vaginal bleeding does not typically occur in appendicitis but would be suggestive of rupture of a corpus luteum cyst, rupture of a graafian follicle, or a ruptured tubal pregnancy.
A 21 years old men comes to ER with left lower quadrant pain. He first noticed that pain early in morning and tells the physician that the pain never be relaxed from that time. At physical examination there is tenderness with palpation on Left lower quadrant and rebaund is possessive. At sonographic examination there is heterogeneous echotexture in mezerenic fat plans and 2 cm bowel wall thickening where the patient's pain the worst. According to these findings what is the most likely clinical diagnosis?

a. Volvulus
b. Diverticulit
c. Ulserative colitis
d. Mezenteric lymphadenopathy
e. Epiploic appendagitis

correct answer: E

The average patient is about 40 years old and develops acute abdominal pain which may be left-sided, right, or central. The pain is sharp and stabbing and may be associated with nausea or vomiting. Fever is usually absent. The symptoms from EA often mimic acute appendicitis, diverticulitis, or cholecystitis. Initial lab studies are usually normal. Typically, a CT scan is ordered to help exclude more serious or surgical problems (above) and the inflammatory changes of EA are seen coincidentally. EA follows a benign, self-limiting course and may be treated with analgesics and expectant observation. The usual time course is about one week. A correct diagnosis is important to avoid unnecessary surgical or medical intervention.
A 35 year-old male with ulcerative colitis presents to his gastroenterologist for one week of jaundice, itching and right upper quadrant pain. Physical exam is notable for icteric sclera, excoriations on the upper shoulder and pain on palpation of the right upper quadrant.

Liver function tests are sent that show:
Total bilirubin 5.3
Direct bilirubin 4.1
AST: 45
ALT: 50
Alkaline Phosphatase: 150

A decision is made for the patient to undergo an ERCP and the cholangiogram is shown above.

Which antibody is most commonly associated with this condition?

a. ASCA (Anti-Saccharomyces cerevisiae antibodies)
b. Anti-mitochondrial antibody
c. Anti-smooth muscle antibody
d. C-ANCA
e. P-ANCA

**Correct answer:** E

This patient is most likely suffering from primary sclerosing cholangitis which is commonly associated with ulcerative colitis. In PSC, the both the intra- and extra-hepatic bile ducts are affected causing strictures. In the cholangiogram this is noted by the alternating strictures and dilated portions of the ducts. In 80% of cases of PSC (and UC) there is an increase in the p-ANCA antibody (option E), although it is not specific for PSC (it is also found in microscopic polyangiitis and Churg-Strauss syndrome among others). The only definitive treatment is liver transplant.

ASCA (option A) is more often correlated with Crohn’s disease.

Antimitochondrial antibody (option B) is correlated with primary biliary cirrhosis. In PBC the intra hepatic bile ducts are affected and therefore the cholangiogram would not show extra hepatic involvement. In addition PBC is more often found in middle aged women.

Anti-smooth muscle antibody (option C) is found in 20-50% of cases of PSC, but is less common than p-ANCA. Just like p-ANCA it is not specific for PSC, it is also found in autoimmune hepatitis and others.

C-ANCA is associated with Wegener’s Disease which affects the lungs and kidneys, but it is not usually associated with hepatic disease.
A 50 year old man presents to his physician with parkinson-like symptoms of resting tremor, rigidity, and bradykinesia. Elements of early dementia are also present. The physician performs a thorough physical examination prior to laboratory and imaging studies and discovers the above finding on slit lamp analysis. The presumptive diagnosis can also present with which of the following findings?

- Decreased fibronectin level
- Decreased ceruloplasmin level
- Increased transferrin level
- Increased haptoglobin level
- Increased albumin level

**Correct answer:** B

This man presents with signs and symptoms of Wilson's disease, also known as hepatolenticular degeneration. This is an autosomal recessive disorder characterized by a defective hepatocyte transport of copper into bile for excretion. As a direct result of this, there is a decrease in synthesis of ceruloplasmin (Choice B), a copper binding protein. Copper then accumulates in various organs, including the liver, brain, kidneys, joints, and eyes. Cirrhosis can be a long term result. Deposition in the brain leads to degeneration of the lenticulate nucleus, accounting for the parkinsonian symptoms. The eye finding of a brown ring surrounding the iris is known as a Kayser-Fleischer ring and represents the deposition of copper around the cornea. As a general rule when a substance accumulates in the body, the binding protein for that substance is decreased and when a substance decreases the binding protein for it is increased in synthesis.

Fibronectin (Choice A) is an ECM glycoprotein that binds integrins. It is not involved in Wilson's disease.

Transferrin (Choice C) is a binding-protein for iron. It is increased in iron-deficiency anemia.

Haptoglobin (Choice D) is a binding-protein for free Hemoglobin. It is decreased in hemolytic anemias, and has been found to be decreased in some presentations of Wilson's disease.

Albumin (Choice E) is the most abundant plasma protein in the blood, and in the setting of a Wilson's disease-induced liver cirrhosis, may actually be decreased.

Image credit: http://upload.wikimedia.org/wikipedia/commons/0/00/Kayser-Fleischer_ring.jpg
A 40 year old woman who is being treated for Crohn’s disease presents with a sudden onset of hemoptysis, low-grade fever, and night sweats of 5 days duration. During the interview, she revealed to you that her “pants feel loose.” Which of the following is the most likely agent she was treated with?

- a. azathioprine
- b. infliximab
- c. methotrexate
- d. prednisone
- e. sulfasalazine

**correct answer: B**

It has been widely accepted that the chronic inflammation seen in Crohn’s disease is due to the increased production of cytokines, notably TNF alpha. These conclusions led to the development of the human-murine chimeric monoclonal antibody against TNF, infliximab (Remicade), which has shown to be effective in treating Crohn’s. Patients taking Infliximab are predisposed to reactivation of latent tuberculosis, which was the most likely explanation of the patient’s symptoms of weight loss, hemoptysis, and night sweats. Hence it is recommended that patients get appropriate screening for TB before taking Infliximab.

Azathioprine is a purine synthesis inhibitor, inhibiting the proliferation of cells, especially leukocytes, also used in the treatment of Crohn’s and autoimmune conditions such as rheumatoid arthritis. Its use does not specifically predispose to TB reactivation.

Methotrexate is a competitive and reversible inhibitor of dihydrofolate reductase (DHFR), an enzyme that participates in the tetrahydrofolate. As folate is important for cell division, reduced folate leads to cell death, which explains the usefulness of this drug in cancer treatment. Its use does not specifically predispose to TB reactivation.

Prednisone is a synthetic corticosteroid that has immunosuppressant effects. Its use does not specifically predispose to TB reactivation.

Sulfasalazine is an anti-inflammatory agent used in the treatment of inflammatory bowel disease synthesis. Its use does not specifically predispose to TB reactivation.
Necrotizing Enterocolitis?

a. ,
b. ,
c. ,
d. ,
e. ,

Correct answer: E

High yield in Step 1, need more Q's on this disease!!!!!!!
Which of the lab test is used for determining the prognosis of acute pancreatitis?

a. Serum Ca++
b. Serum Amylase
c. Urinary Amylase
d. Serum Lipase
e. TLC

**Correct answer:** A

Because serum Ca++ will be low indication soap formation i.e Necrosis. so more hypocalcemia means more necrosis and hence soaponification.
A 55-year-old man presents to a physician with jaundice. Ultrasonography demonstrates a 5 cm mass in the head of the pancreas. Endoscopic retrograde cholangiopancreatography with cytologic sampling demonstrates cells with large hyperchromatic nuclei and a high nuclear/cytoplasmic ratio. A few small glands composed of these cells are also seen in the cytologic preparation. The overall prognosis for this man will be most similar to that of a patient with which of the following malignancies

a. Adenocarcinoma of the breast  
b. Adenocarcinoma of the colon  
c. Adenocarcinoma of the esophagus  
d. Adenocarcinoma of the prostate  
e. Primary gastric lymphoma

**correct answer:** C

The patient probably has pancreatic adenocarcinoma. This cancer carries one of the worst prognoses, with a 3.5% overall 5 year survival rate despite all attempts at aggressive management. The prognosis is also bleak with adenocarcinoma of the esophagus, with a 10% overall 5 year survival rate.

Adenocarcinoma of the breast (choice A) now has an overall 5 year survival rate of 60-70%.

Adenocarcinoma of the colon (choice B) now has an overall 5 year survival rate of 50-60%.

Adenocarcinoma of the prostate (choice D) now has an overall 5 year survival rate of 50-70%.

Primary gastric lymphoma (choice E) has an overall survival 5 year survival rate of 75-85%.
An afebrile 25-year-old male who recently emigrated from Eastern Europe presents with painless enlargement of the left parotid gland over the last six months. You feel a firm, movable mass. He has never smoked or consumed alcohol. He did have significant dental work (x-rays and surgery) on that side as a child. If you were to biopsy the gland, what would you expect to see microscopically?

a. A poorly encapsulated cribriform mass of glands and hyaline material
b. A poorly-encapsulated mass of squamous, mucoid, and intermediate cells
c. A well-encapsulated mass of myoepithelial tissue in a mesenchymal matrix
d. An encapsulated mass of cystic spaces, papillary epithelium, and germinal centers
e. Lymphocytic infiltrate

**Correct answer:** B

This patient has mucoepidermoid carcinoma, a painless, slow-growing malignant neoplasm, usually found as a firm mass in the parotid gland. It is commonly caused by radiation, which is suggested by his history of dental work in an area of the world where excessive amounts of radiation might be used. Therefore, on histological examination you would expect to find a poorly-encapsulated mass of squamous, mucoid, and intermediate cells.

A -- A poorly encapsulated cribriform mass of glands and hyaline material suggests an adenoid cystic carcinoma. These malignant neoplasms are most common in the salivary glands of the palate, where they grow painfully and tend to invade nerves. This patient's tumor is not in the palate.

C -- A well-encapsulated mass of myoepithelial tissue in a mesenchymal matrix suggests pleomorphic carcinoma, a slow-growing, benign neoplasm. It is the most common salivary gland tumor. The patient's history of radiation exposure makes B the better answer.

D -- An encapsulated mass of cystic spaces, papillary epithelium, and germinal centers suggests Papillary Cystadenoma Lymphomatosum, or Warthin's Tumor. This benign neoplasm is probably heterotopic salivary tissue trapped in a regional lymph node. It is most commonly found in male smokers, but this patient has never smoked.

E -- Lymphocytic infiltrate is suggestive of an infection, such as Mumps, caused by a paramyxovirus. Mumps is more dangerous in older patients than in children, and you could be concerned about this patient developing orchitis and infertility if he has not had all his immunizations. However, the parotitis of Mumps is bilateral 90% of the time, and patients usually develop a fever. There is a better answer.
A 65-year-old Caucasian female presents with headaches and back pain of two months duration. A radiograph of her skull reveals "punched-out" osteolytic lesions. You suspect a hematologic malignancy. Which of the following findings are also likely?

I. Erythrocytic Rouleaux formations
II. Reed-Sternberg cells
III. Plasma cell proliferation in bone marrow

a. I only  
b. II only  
c. III only  
d. I & II  
e. I & III

**Correct answer: E**

The likely diagnosis is multiple myeloma. The disease is characterized by overpopulation of neoplastic plasma cells in the bone marrow, whose hypersecretion of antibody results in several circulatory abnormalities, among them string-like aggregations of erythrocytes. Other than the most common complaint of bone pain, common presentations for multiple myeloma include pathologic fractures (a fracture secondary to minor trauma), bone marrow suppression (from myelophthisis) resulting in anemia, renal failure (due to either hypercalcemia or the increased protein load - Bence-Jones proteins may be seen in the urine), and neurological symptoms due to hypercalcemia. Reed-Sternberg cells are seen in Hodgkin's Lymphoma.

Note: There are no questions of this format on USMLE Step I.
A 75-year-old man presents complaining of progressively worsening fatigue and weight loss. He has lost 30 lbs over the past two months and is losing his appetite. A stool guaiac is positive and his hematocrit is 32%. Considering the most likely cause of this patient's anemia, which value is most likely to be decreased?

a. RBC protoporphyrin  
b. Serum ferritin  
c. Serum iron  
d. Soluble transferrin receptor levels  
e. Total iron binding capacity

**Correct answer:** C

This patient likely has an iron-deficiency anemia secondary to occult blood loss (possibly due to GI bleeding secondary to NSAIDS, given the positive stool guiac and a history of arthritis). The stages of iron deficiency in sequence are as follows: absent iron stores; decreased serum ferritin; decreased serum iron; increased TIBC; decreased iron saturation(%); normochromic normocytic anemia; microcytic hypochromic anemia. The total iron binding capacity will be elevated as there are relatively "more" binding sites for iron on transferrin receptors. Increased transferrin levels will also increase the soluble receptor levels. RBC protoporphyrin will rise as iron is needed to proceed from this intermediate in heme synthesis.

Serum ferritin in iron-deficiency anemia is generally low, as it reflects the iron "stores" in the bone marrow (which will be depleted). However, in patients with concomitant inflammatory states such as cancer (possible colon cancer in this patient) or autoimmune disease (rheumatoid arthritis in this patient), ferritin levels may not be decreased as it is also an acute phase reactant.

S. Iron & S. transferrin are used in suspected iron deficiency with concomitant inflammatory disease or cancer. However even these are acute phase reactants and may not be decreased. Recently, the serum transferrin receptor (sTfR) to ferritin ratio has been used to diagnose iron deficiency anemia in the setting of a high or normal ferritin, but the sTfR isn't widely available. The measurement of serum transferrin receptor (TfR) is proving to be very sensitive in identifying iron deficiency in problematic patients, including the elderly with chronic diseases and possibly pregnant women.

The most definitive test of iron deficiency is a bone marrow biopsy. The bone marrow is part of the iron storage pool, and if a patient is truly iron-deficient, it will contain no stainable iron. If your patient can't or won't have a bone marrow biopsy, a careful therapeutic trial of iron can help clinch the diagnosis.
A 3 year old presents to your clinic with his mother. Upon examination, you discover a painless flank mass in the abdominal area. His mother also complains her son has had blood in his urine. What chromosome is associated with this disease?

a. Chromosome 11  
b. Chromosome 13  
c. Chromosome 21  
d. Chromosome 18  
e. Chromosome 16

**Correct answer:** A

This young child presents with the most common renal malignancy of childhood, a Wilms Tumor. This can also be apart of the WAGR complex, Wt-1 tumor suppressor gene, and hemihypertrophy of the body.
A middle age patient presents with history of tiredness, pain in his abdomen, and sexual dysfunction. Blood lab results shows a decrease serum haptoglobin, and a normocytic anemia with pancytopenia. Sucrose hemolysis test was positive. The patient's condition is best described as

a. Intrinsic hemolytic anemia/extravascular hemolysis
b. Extrinsic hemolytic anemia/intravascular hemolysis
c. Intrinsic hemolytic anemia/intravascular hemolysis
d. Extrinsic hemolytic anemia/extravascular hemolysis
e. Positive Direct Coomb’s Test/extravascular hemolysis

correct answer: C
C is the correct answer
The patient most likely suffers from paroxysmal nocturnal hemoglobinuria, an acquired stem-cell disorder characterized by complement-induced hemolytic anemia, red urine (due to the appearance of hemoglobin in the urine) and thrombosis (in this case in either the superior or inferior mesenteric vein causing ischemia). Erectile dysfunction is due to rapid destruction of the RBCs.

A IS INCORRECT: Intrinsic hemolytic anemia/extravascular hemolysis most likely the case of sickle cell anemia.

B IS INCORRECT: Extrinsic hemolytic anemia/intravascular hemolysis most likely the case of macroangiopathic hemolytic anemia (aortic stenosis, is the most common cause).

D IS INCORRECT: Extrinsic hemolytic anemia/extravascular hemolysis most likely the case of warm autoimmune hemolytic anemia.

E IS INCORRECT: Positive Direct Coomb’s Test/extravascular hemolysis most likely the case of drug-induced hemolytic anemia.
A middle age patient presents with history of tiredness, pain in his abdomen, and sexual dysfunction. Blood lab results shows a decrease in serum haptoglobin, and a normocytic anemia with pancytopenia. Sucrose hemolysis test was positive. The patient’s condition is most likely attributed to which of the following

- a. Intrinsic hemolytic anemia/extravascular hemolysis
- b. Extrinsic hemolytic anemia/intravascular hemolysis
- c. Intrinsic hemolytic anemia/intravascular hemolysis
- d. Extrinsic hemolytic anemia/extravascular hemolysis
- e. Positive Direct Coomb’s Test/extravascular hemolysis

**Correct answer:** C

C is the correct answer.

Serum haptoglobin is a protein that scavenges intravascular Hb. So if it reduced it means that a intravascular hemolytic process is occurring. Also in intravascular hemolysis we will see hemoglobinuria. The patient most likely suffers from paroxysmal nocturnal hemoglobinuria, an acquired stem-cell disorder characterized by complement-induced hemolytic anemia red urine (due to the appearance of hemoglobin in the urine) and thrombosis (in this case in either the superior or inferior mesenteric vein causing ischemia). Erectile dysfunction is due to rapid destruction of the RBCs. Screening test for PNH is sucrose hemolysis test or sugar water test in which sucrose enhances the complement mediated destruction of RBC’s. Confirmatory test is Ham test in which acidified serum activates alternate pathway of complement causing RBC lysis.

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E IS INCORRECT: Positive Direct Coomb’s Test/extravascular hemolysis most likely the case of drug-induced hemolytic anemia.
A 30 year old woman presents to her physician with a 1 month history of night sweats, weight loss, and low grade fever. On exam, she is found to have nontender cervical and supraclavicular lymphadenopathy. A peripheral blood smear reveals numerous cells identical to the one pictured above. The doctor reveals the diagnosis to the patient, and tells her that it occurs most commonly in young women. Which of the following is the most likely disease process present in this patient?

a. Hodgkin's lymphoma: lymphocyte predominant subtype  
b. Hodgkin's lymphoma: mixed cellularity subtype  
c. Hodgkin's lymphoma: nodular sclerosing subtype  
d. Non-Hodgkin's lymphoma: follicular (small cleaved cell) subtype  
e. Non-Hodgkin's lymphoma: lymphoblastic lymphoma subtype  

**Correct answer:** C

This woman has the most common subtype of Hodgkin's lymphoma (HL) known as nodular sclerosing. It accounts for 65-75% of HL and unlike the other subtypes (which occur most commonly in men), occurs primarily in women. Its pathology reveals collagen banding separating nodular regions. HL, which comprises ~40% of all adult lymphomas, is overall characterized by painless lymphadenopathy often involving a single group of nodes, the presence or absence of constitutional "B" symptoms, and the characteristic Reed Sternberg (RS) cell. This binucleate "owl eyed" cell, which is of B cell origin, is pathognomonic for HL, occurring in all the subtypes to varying degrees. A better prognosis is suggested by fewer RS cells. There is a close link between HL and the presence of EBV in the bloodstream.

The lymphocytic predominant subtype (choice A) makes up 5-10% of Hodgkin's diagnoses. It is pathologically characterized by an abundance of lymphocytes relative to RS cells. It occurs most often in men under 35 years old.

The mixed cellularity subtype (choice B) makes up 25% of Hodgkin's diagnoses. It is pathologically characterized by a mixture of lymphocytes and RS cells, and is the subtype often noted to have the greatest abundance of RS cells. It has the closest EBV association among all the subtypes.

The follicular (small cleaved cell) subtype (choice D) is the most common indolent Non-Hodgkin's lymphoma (NHL). Pathologically, it attempts to mimic lymph node architecture by forming follicles. Its growth can be tied directly to its genetic link; it involves overexpression of bcl-2, an inhibitor of apoptosis. NHL, which comprises 60% of adult lymphomas, is overall characterized by painless lymphadenopathy often involving multiple sets of lymph nodes, fewer constitutional "B" symptoms, and a predominantly B cell origin. Immunosuppressed states are a strong risk factor for the development of NHL.

The lymphoblastic lymphoma subtype of NHL is the only subtype with a T cell origin. It presents most often in children with an accompanying mediastinal mass.

Image credit: http://library.med.utah.edu/WebPath/jpeg5/HEME045.jpg
A 43 year old man presents to his physician, with complaints of enlargement of right palatine tonsil over the last several months. The tonsil is nontender on physical examination, biopsy of the affected tonsil reveals complete effacement of germinal centers, which are replaced by irregular nodules composed of small lymphocytes. Which of the following is the most likely diagnosis.

- a. Diffuse Lymphoma
- b. Follicular Lymphoma
- c. Hodgking Disease
- d. Reactive Lymphoid Hyperplasia
- e. Tonsilar abscess

**Correct answer:** B

B. is correct; the tonsil is composed of lymphoid tissue, and can be affected by any process that involves lymph nodes. The description of many irregular nodules with effacement of germinal centers by small lymphocytes specifically suggests a follicular (nodular) lymphoma. All follicular lymphomas are derived from B-cell lineage.

A description of diffuse lymphoma (choice A) would mention sheets of lymphocytes without nodules.

A description of Hodgking disease (choice C) would probably include the Reed sternberg cells and eosinophils.

A description of reactive lymphoid hyperplasia (choice D) would probably include many germinal centers.

A description of tonsilar abscess (choice E) would include neutrophils and necrosis.
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1. In a patient with deficiency in Von Willebrand factor, what abnormalities are you likely to observe?

<table>
<thead>
<tr>
<th>Bleeding time</th>
<th>Platelet time</th>
<th>half life of Factor VIII</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Increased</td>
<td>Increased</td>
<td>Decreased</td>
</tr>
<tr>
<td>B. Increased</td>
<td>Normal</td>
<td>Decreased</td>
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<tr>
<td>C. Decreased</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>D. Decreased</td>
<td>Normal</td>
<td>Increased</td>
</tr>
<tr>
<td>E. Increased</td>
<td>Normal</td>
<td>Normal</td>
</tr>
</tbody>
</table>

Correct answer: B

a. A  
b. B  
c. C  
d. D  
e. E
A 60-year-old woman comes to a primary care physician for the first time in 30 years. She has experienced significant weight-loss recently (20 lbs in 2 months) and is thought to have developed cancer (diagnosis awaiting biopsy results). Which of the following conditions is a precursor to cancer and could have been detected on routine clinical examination?

a. Actinic keratosis  
b. Adrenal adenoma  
c. Fibrocystic breast disease  
d. Transitional cell carcinoma  
e. Uterine leiomyoma

**Correct answer:** A

Actinic keratosis is a pre-malignant condition to squamous cell carcinoma of exposed areas of skin. It results from progressive dysplastic changes that begin in the basal layer of the epidermis. The most important risk factor is chronic exposure to UV light. If removed early, SCC can safely be prevented. Fibrocystic breast changes are benign and occur in a cyclical pattern with menstruation. Uterine leiomyomas, aka "fibroids" may cause bleeding but are benign. Transitional cell carcinoma, or cancer of the bladder wall, is rare and not routinely screened for. Adrenal adenomas are also not routinely screened for, and are often discovered incidentally by CT for other reasons.
A 76 year-old woman with a long history of rheumatoid arthritis presents complaining of weakness and swelling of her hands and feet. On exam, BP is 140/78, HR is 76, and RR is 12. There is mild hepatomegaly. Urinalysis shows proteinuria and liver function tests are mildly abnormal. Congo red stain of liver biopsy shows apple green birefringence when viewed under polarized light. Accumulation of which protein accounts for this finding?

a. Amyloid Associated protein  
b. Amyloid Lightchain protein  
c. Amylin  
d. Collagen  
e. Transthyretin

_correct answer:_ A

This patient has secondary amyloidosis due to long-standing rheumatoid arthritis, which results in accumulation of AA protein. Other causes of secondary amyloidosis include tuberculosis, osteomyelitis, and syphilis. AL protein is seen in primary amyloidosis. Transthyretin is seen in senile amyloidosis. Amylin is seen in amyloidosis secondary to type II diabetes mellitus.
A 70 year-old woman with a family history of breast cancer has a lumpectomy after a breast mass is identified during screening mammography. If pathological, histological, and cytogenetic studies were performed, which of the following would be most suggestive of adenocarcinoma?

a. Desmoplastic surrounding tissues  
b. Encapsulated tumor  
c. Polyclonality  
d. Predominantly interphase cells  
e. Well-differentiated cells

correct answer: A

Adenocarcinoma is commonly associated with desmoplasia, a tumor-induced proliferation of non-neoplastic fibrous connective tissue which surrounds the tumor. It is seen most commonly in adenocarcinoma of the breast, pancreas, and prostate.

Well-differentiated cells (E) and encapsulation of the tumor (B) would be more indicative of benign neoplasms than adenocarcinoma, which is malignant. Polyclonality (C) is incorrect because neoplasms, both benign and malignant, are monoclonal. A predomination of interphase cells (D) would indicate controlled cell division, while adenocarcinomas undergo rapid cell division (and thus have a high mitotic index).
A 45 year old woman presents with fevers and weight loss. A malignancy workup yields evidence of disseminated cancer. A biopsy of the likely source shows concentric laminar calcified collections resembling "Orphan Annie eyes." Which of the following tumors was the likely source of the biopsy?

a. Mucinous cystadenocarcinoma of ovary
b. Glioblastoma multiforme
c. Invasive ductal carcinoma of breast
d. Adenocarcinoma of lung
e. Papillary adenocarcinoma of thyroid

**Correct answer: E**

"Orphan Annie eye" is the classic description of nuclear inclusions found in papillary adenocarcinoma of the thyroid (Answer E). Also present in papillary thyroid carcinoma are lamellar calcifications known as psammoma bodies, which also can be found in serous papillary cystadenocarcinoma of the ovary, meningiomas, and malignant mesothelioma.

Mucous cystadenocarcinoma of the ovary, as the name suggests, produces a lot of mucus, which can lead to pseudomyxoma peritonei. Glioblastoma multiforme, the most common type of adult primary brain tumor, is known to have "pseudopalisading" tumor cells. This means the tumor cells border central areas of necrosis and hemorrhage. Invasive ductal carcinoma of the breast shows ductal tissue that invades surrounding breast tissue. Adenocarcinoma of the lung is the most common cause of lung cancer in non-smokers. It appears as abnormal glandular tissue invading surrounding lung tissue.
A 65-year-old man with a history of mitral valve prolapse presents with fever and weight loss after a recent dental procedure. At one week in the hospital, he has acute left flank pain. A CT shows a wedge-shaped area of hypoperfusion in the left kidney. If a biopsy of this area were done, what would it show?

- Atrophy
- Caseous necrosis
- Coagulative necrosis
- Dysplasia
- Liquefactive necrosis

**Correct answer:** C

The man presumably has bacterial endocarditis (likely Strep viridans) that occurred when oral flora entered his bloodstream during a dental procedure and infected his abnormal heart valve. A complication of endocarditis is systemic septic emboli, often to the spleen or kidney. Tissue death due to hypoperfusion is called coagulative necrosis.

An exception is hypoperfusion to brain tissue, in which for unknown reasons, dead tissue is dissolved, a process called liquefactive necrosis (choice E).

Caseous necrosis (choice B) is most often associated with tuberculous granulomas, appearing "cheese-like" on microscopy.

Atrophy (choice A) is shrinkage of normal tissue, as occurs with unused muscle tissue.

Dysplasia (choice D) refers to abnormal changes in cell architecture that occurs in pre-malignant or malignant tumors. Generally, dysplasia indicates proliferation of immature cells with a corresponding decrease in the number of mature cells.
A pathology resident discovers that the cheap labels on a box of pathology slides have peeled off. He looks at one slide under the microscope and compares it to the one-line patient summaries he has been given. The slide shows fat necrosis. To whom does this slide most likely belong?

- a. 4-year-old boy with liver abscess
- b. 33-year-old woman with pulmonary tuberculosis
- c. 48-year-old man with myocardial infarction
- d. 55-year-old alcoholic man with acute pancreatitis
- e. 86-year-old woman with stroke

correct answer: D

When pancreatic cells are injured during acute inflammation (pancreatitis), they release lipases that digest surrounding fat into soaps, a process called fat necrosis (Answer D). Tuberculosis is characterized by caseous necrosis; granulomas that look like cheese. Abscesses are collections of liquefied dead tissue and neutrophils, which is known as liquefactive necrosis. Cell death due to hypoperfusion, as in MI, is called coagulative necrosis. The exception is when brain tissue dies, because it is the only tissue that undergoes liquefactive necrosis.
A 5-week-old girl, who appeared to be healthy at birth, develops diarrhea and vomiting a few days after. Your current examination reveals that she has hepatomegaly, jaundice, and early cataract formation and is not meeting developmental milestones. The most likely diagnosis is:

a. Galactosemia
b. Hurler syndrome
c. Pyloric stenosis
d. Tay-Sachs disease
e. Type I glycogenosis

correct answer: A

Galactosemia is an autosomal recessive disorder due (in this more common and more severe form of the disease) to a lack of galactose-1-phosphate uridyl transferase. This results in the formation and accumulation of galactose metabolites. If the infant’s diet is not modified to exclude milk products, this will result in damage to the liver (fatty change, cholestasis, cirrhosis, liver failure), eyes (cataract formation), and brain (mental retardation).

Hurler syndrome (choice B) is a severe form of mucopolysaccharidosis that typically becomes apparent between 6 months and 2 years of age. Caused by deficiency of enzyme L-Iduronidase. Prominent features include coarse facies, dwarfism, organomegaly, cataracts, and mental retardation, not diarrhea, vomiting, and jaundice.

Pyloric stenosis (choice C) can occur as a congenital condition, more frequently in baby boys (M:F = 4:1). It is marked by projectile vomiting in the first month of life, but not the other findings in this case. Tay-Sachs disease (choice D) is a lipid storage disease due to a deficiency of hexosaminidase A. There is an inexorable deterioration of mental and motor functions within a few months of birth culminating in a vegetative state and death within 3 or 4 years. Type I glycogen storage disease or von Gierke disease (choice E) is due to a deficiency of glucose-6-phosphatase and usually becomes apparent in the first year of life as hypoglycemia and/or hepatomegaly.
34 years old women comes to the clinic a few hours after being treated for a Louse carried disease, her current symptoms consists of fever of 102.7F, exacerbation of skin lesions, myalgia and thumbling headache. What can be given to treat these side effects?

- a. Aspirin
- b. Ofloxacin
- c. Metronidazole
- d. Oprelvekin
- e. Tenofovir

**Correct answer:** A

Herxheimer reaction happens for all the Spirochaete’s, and is due to the necrotic lipopolysaccharides. Typically the death of these bacteria and the associated release of endotoxins occurs faster than the body can remove the toxins via the natural detoxification process performed by the kidneys and liver. It is manifested by fever, chills, headache, myalgia (muscle pain), and exacerbation of skin lesions. Duration in syphilis is normally only a few hours but can be much longer, up to months or years, for other diseases. The intensity of the reaction reflects the intensity of inflammation present. In Relapsing Fevers (tick and louse borne) it usually begins within a few hours of the first dose and causes an initial rise in temperature, pulse rate and blood pressure, then followed by marked vasodilation & sweating, which can result in shock. The Herxheimer reaction has shown an increase in inflammatory cytokines during the period of exacerbation, including tumor necrosis factor alpha, interleukin-6 and interleukin-8.

- B) DNA replication inhibitor, and it interferes with bacterial DNA topoismerase II & IV
- C) Bactericidal, interferes with nucleic acid synthesis
- D) IL-11 agonist, Increases platlet formation used in thrombocytopenia
- E) Protease inhibitor which can cause Fanconi Syndrome.
Researchers are trying to determine genetic influences on myocardial infarction survival. Various levels of cholesterol are injected daily into a population of genetically diverse mice populations. Mice are autopsied immediately upon death. Which of the following finding on myocardial tissue biopsy would represent reversible cellular damage?

- a. Increased cellular sodium and cellular edema
- b. Increased cellular Calcium
- c. Increased cellular pH
- d. Increased cellular cytochrome C
- e. karyolysis

**Correct answer:** A

Cellular edema due to an interruption of the Na/K pump is the only choice that represents reversible damage.

All other answer choices are found in irreversible cell damage.
Tumors can be classified by both grading and staging. The difference between these two is that tumor grading tends to be based on _______ while tumor staging tends to be a measure of the degree of _______.

a. Differentiation, Spread  
b. Differentiation, Treatability  
c. Spread, Differentiation  
d. Spread, Treatability  
e. Treatability, Spread

**correct answer:** A

Tumor grading is I - IV and is based on differentiation.

Tumor staging typically utilizes the TNM staging system and is based on spread.

T- Tumor size  
N- lymph Nodes  
M- Metastases
The "Philadelphia Chromosome" results from a reciprocal translocation \([ t(9:22) ]\) and causes Chronic Myelogenous Leukemia (CML). Gleevec treats this by binding to the oncogene formed by this translocation, namely:

a. Bcr-abl  
b. C-kit  
c. C-myc  
d. Erb-B2  
e. Ras

**Correct answer:** A

BCR-ABL is the oncogene formed by this translocation.

Note: C-myc is formed in Burkitt's Lymphoma \([ t(8:14) ]\)
A 53-year-old man presents to the emergency room with fever, complaining of several weeks of lower thoracic spinal pain, erythema, and swelling. Chest x-ray reveals loss of intervertebral space with vertebral collapse, as well as a cavitary lesion in the right apical lung field.

What is the most likely pathology of his complaint?

a. Decreased osteoblast activity with normal osteoclast activity  
b. Haemophilus influenzae via hematogenous spread  
c. Mycobacterium tuberculosis via direct extension  
d. Mycobacterium tuberculosis via hematogenous spread  
e. Type I collagen defect

**Correct answer:** D

This patient is suffering from tuberculous osteomyelitis of the spine (Pott’s disease). Although these patients often present with a clear chest x-ray, pulmonary lesions suggestive of tuberculosis help confirm the diagnosis. Lumbar and lower thoracic vertebrae are common sites of hematogenous targets, owing to their dense vascularization, making Mycobacterium tuberculosis via hematogenous spread (choice D) the correct answer.

Decreased osteoblast activity with normal osteoclast activity (choice A) is seen in primary osteoporosis type 2 (“senile osteoporosis”), which occurs in patients over 60 of both genders.

Haemophilus influenzae via hematogenous spread (choice B) is a potential cause of vertebral osteomyelitis, it is unlikely in a patient with concomitant tuberculosis.

Mycobacterium tuberculosis via direct extension (choice C) is unlikely, as it would require the lesion to spread through lung parenchyma, into the pleural space, and through the parietal pleura. The discontinuous nature of the pulmonary and vertebral lesions (apical vs. low thoracic) further discounts this possibility.

A type I collagen defect (choice E) would be seen with genetic disorders like osteogenesis imperfecta (OI). Ehlers-Danlos Syndrome (EDS) HAS TYPE 3 COLLAGEN DEFECT. In OI, fractures are usually found in the long bones, ribs, and small bones of the hands and feet. The classic symptom of EDS is joint laxity, manifested by dislocations and subluxations. Neither disorder would present with signs of infection.
A 32 year old female complains of difficulty getting up from the toilet for the past 6 weeks accompanied by soreness of her leg muscles. She has never experienced similar symptoms in the past and has no other complaints. Physical exam is significant for decreased strength in her thighs bilaterally, with reflexes intact, and a painless purple discoloration above her eyelids bilaterally (see picture). A muscle biopsy is performed. What would the pathology likely reveal?

a. Dense eosinophilic inclusions
b. Increased variability of fiber size with atrophy and hypertrophy
c. PAS-positive staining of clear vacuoles within myocytes
d. Perimysial inflammation with lymphocytic infiltrate
e. Ragged red fibers on trichrome stain

**Correct answer:** D

Difficulty rising from a toilet is indicative of proximal muscle weakness; together with the typical heliotrope rash, this finding leads to a diagnosis of dermatomyositis, which should also prompt a workup for an underlying malignancy or systemic disease. The pathological findings in dermatomyositis include perimysial inflammation with lymphocytic infiltrate (Choice D).

Dense eosinophilic inclusions (Choice A) are typical of inclusion body myositis.

Increased variability of fiber size with atrophy and hypertrophy (Choice B) is typical of Becker’s muscular dystrophy.

PAS-positive staining of clear vacuoles within myocytes (Choice C) would be seen in a glycogen storage disease.

In patients with mitochondrial muscle disease, ragged red fibers are seen on trichrome stain (Choice E).
A 10-year old boy presents to his physician with pain and swelling in the left femur. Onion-skin lytic lesions were seen on radiological imaging. On histological examination of the lesion, anaplastic blue small cells were seen. What chromosomal translocation would result in such a clinical picture?

a. t(14;18)  
b. t(8;14)  
c. t(9;22)  
d. t(15;17)  
e. t(11;22)

**Correct answer:** E
The clinical picture is that of Ewing's Sarcoma which is associated with t(11;22) (choice E).

Choice A is associated with follicular lymphoma; the gene involved is Bcl-2.  
Choice B is associated with Burkitt's lymphoma; the gene involved is c-myc.  
Choice C is associated with chronic myelogenous leukemia; the gene involved is BCR-ABL. Note that this is the target of a drug - imatinib (Gleevec).  
Choice D is associated with acute myelogenous leukemia.
question templates
a. a
b. b
c. c
d. d
e. e

correct answer: B
A 42-year-old man is rushed to the emergency room by his wife after she comes home and cannot rouse him successfully from sleep. He has been sick for approximately three days with a reported temperature of 102°F. On exam the patient is somnolent and his face appears asymmetrical with his left eyelid and cheek drooping in comparison to the right side. His temperature is 103°F. The patient undergoes an MRI scan as seen. What is the most likely agent responsible for this patient’s symptoms?

- Coxsackie virus
- Herpes Simplex Type I (HSV-1)
- Mycobacterium tuberculosis
- Neisseria meningitidis
- Streptococcus pneumoniae

**Correct answer:** B

This patient is suffering from encephalitis from infection with HSV-1 resulting in characteristic hemorrhagic necrosis of the temporal lobes that can be seen on the MRI. All of the other answer choices typically cause meningitis. Encephalitis refers to infection of the brain tissue and results in a clinical syndrome often characterized by the rapid onset of fever, headache, seizures, focal neurologic signs, such as the facial nerve palsy displayed by this patient, and impaired consciousness which may proceed to coma. Meningitis refers to inflammation of the leptomeninges, the tissues surrounding the brain and spinal cord. The presence or absence of normal brain function is the distinguishing feature between encephalitis and meningitis. Patients with meningitis may be uncomfortable, but their cerebral function remains intact. Encephalitic patients, however, frequently display abnormal brain function, including altered mental status, motor or sensory deficits, altered behavior and personality changes, and speech or movement disorders.

A. Coxsackie virus is an enterovirus. Enteroviruses are the most common cause of viral meningitis.

C. Mycobacterium tuberculosis is a rare cause of meningitis. Patients may present with protracted headache, vomiting, confusion and cranial nerve signs.

D. Neisseria meningitidis is the second most common cause of bacterial meningitis in adults after S. pneumoniae.

E. S. pneumoniae is the most common cause of bacterial meningitis in adults.
A 6 year old child is brought into your clinic for evaluation of right hand weakness. The mother is unsure of the onset of the weakness, but has been noticing that her child rarely uses his right hand and that the right arm looks unusually limp. Physical exam reveals atrophy of both the thenar and hypothenar eminences, atrophy of the interosseous muscles, and sensory deficits on the medial side of the forearm and hand. Which of the following is the most likely location of the pathologic finding in this patient?

a. Carpal tunnel  
b. Medial epicondyle  
c. Shaft of humerus  
d. Supracondyle of humerus  
e. Costoclavicular space

**Correct answer: E**

Thoracic outlet syndrome (TOS) is most likely to explain all the neurologic findings in this patient. TOS can be present as a congenital anomaly of the costoclavicular space (choice E) which is bordered by the clavicle, first rib, and the superior margin of the scapula. The subclavian vessels and the lower trunk of the brachial plexus (C8, T1) pass through this space and can be compressed by anatomic anomalies such as congenital bands, supernumerary ribs, cervical ribs, or laxity of the costoclavicular joint. An additional finding may include the disappearance of the radial pulse when the patient turns his head to the opposite side.

The carpal tunnel (choice A) and the supracondyle of the humerus (choice D) are common sites of injury to the median nerve. Median nerve damage does not explain hypothenar atrophy, interosseous atrophy, or numbness to the medial side of the forearm and hand, all of which are signs of ulnar nerve involvement. Therefore, the location of pathology is more proximal.

The medial epicondyle (choice B) is a common site of injury to the ulnar nerve, which may explain most of the neurologic findings except for thenar atrophy which suggests median nerve involvement. Therefore, the location of pathology is more proximal.

The shaft of the humerus (choice C) is a common site of injury to the radial nerve which is not implicated in this scenario.
A 40 year old man presents to your clinic for evaluation of progressive loss of sensation in both his arms. Further questioning reveals that the patient was in a motor vehicle accident a few years ago and suffered a cervical spinal cord injury. He has since made a full recovery without any neurological symptoms until now. Physical exam reveals loss of pain and temperature sensation bilaterally, but preservation of vibratory sense and muscle strength. A sagittal MRI T1 image of the patient's cervical spine is shown. This finding on MRI is commonly associated with which of the following conditions?

a. Pseudotumor cerebri
b. Dandy-Walker malformation
c. Arnold-Chiari malformation
d. Alzheimer's disease
e. Scoliosis

correct answer: C

This patient has syringomyelia of the cervical spinal cord secondary to cervical spinal cord injury. Syringomyelia is a degenerative disease of the spinal cord that is characterized by a fluid-filled cavity within the cervical spinal cord. This condition can complicate up to 4% of spinal cord injury and can take months to years to develop. The cavity can continue to expand and eventually compress the anterior white commissure of the cervical spinal cord which contains the crossing fibers of the lateral spinothalamic tracts. The lateral spinothalamic tract carries fibers responsible for pain and thermal sensation. Syringomyelia has been associated with Arnold-Chiari malformation (choice C).

Pseudotumor cerebri (choice A) is a condition characterized by increased intracranial pressure in the absence of tumor and obstruction to CSF flow. CT and MRI are usually normal.

Dandy-Walker malformation (choice B) is a condition characterized by hypoplasia of the cerebellar vermis and cystic dilation of the fourth ventricle.

Alzheimer's disease (choice D) is associated with the radiologic finding of hydrocephalus ex vacuo which describes a dilated appearance of the ventricles when the brain mass is decreased.
A 32-year-old woman with no significant medical history presents to the clinic with visual complaints. On a recent trip to Arizona, she suffered an acute episode of visual impairment. She experienced blurred and double vision, which made it difficult for her to drive. On visual examination, her vision is 20/20 in both eyes. When she is asked to look to the right, her left eye only reaches midline while her right eye shows a beating nystagmus. Testing for visual convergence is intact. Based on the clinical history and physical examination, what is the most likely cause of her complaints?

a. Amaurosis fugax  
b. Cataract  
c. Multiple sclerosis  
d. Optic neuritis  
e. Panophthalmitis

**Correct answer: C**

The patient's history is consistent with a lesion in the medial longitudinal fasciculus (MLF). The MLF connects the nuclei of CNIII (oculomotor) and CNVI (abducens) to facilitate coordination of saccadic eye movements. Lesions in the MLF prevent this coordination from occurring, causing the adducting eye to move only to the midline, while the abducting eye will have nystagmus towards the adducting eye (towards its own midline). Demyelinating lesions in the MLF are a common early manifestation of multiple sclerosis (especially in a young woman with no other disease processes).

(Choice A) Amaurosis fugax is a transient monocular vision loss, generally caused by a thrombus or emboli. The patient has 20/20 acuity, which makes this unlikely.

(Choice B) Cataract is a gradual opacification of the lens of the eye. It does not present acutely as in this patient, and acuity is affected.

(Choice D) Optic neuritis is an inflammation of the optic nerve, which is the most common presentation of multiple sclerosis in young women. Presentation is acute, often painful loss, partial loss of visual acuity, and often color vision is affected. This patient should be evaluated for optic neuritis, but her presentation does not suggest it.

(Choice E) Panophthalmitis is an acute, suppurative (neutrophil-dense) often overwhelming inflammation of the inner eye tissue and sclerae.
A 28-year-old G2P1 with gestational diabetes gives birth to a 4490 g baby boy at 40 weeks. The delivery was complicated by shoulder presentation, but the baby was delivered using the McRoberts maneuver and suprapubic pressure. On exam, the baby has a strong cry, but is found to have asymmetric arm movements. On further exam, the baby’s left arm is adducted and internally rotated. Which of the following is the most likely diagnosis?

a. Clavicular fracture
b. Diaphragmatic paralysis
c. Erb’s palsy
d. Humerus fracture
e. Klumpke’s palsy

**Correct answer:** C

The correct answer is Erb’s Palsy. This is the most likely diagnosis given the above physical exam and risk factors, including maternal gestational diabetes and macrosomia. Erb’s palsy is a C5 and C6 injury that accounts for the majority of obstetrical brachial plexus nerve palsies in newborns. The etiology of these nerve palsies may be iatrogenic lateral traction on the fetal head, usually during deliveries in which there is shoulder dystocia. Arm weakness involves the deltoid and infraspinatus muscles (mainly C5) and biceps (mainly C6). On physical exam, the upper arm is adducted and internally rotated with the forearm extended, but the hand and wrist are spared.

Klumpke’s palsy (C8-T1 nerve injury) is not correct since this is seen much less frequently and often presents with hand paralysis and ipsilateral Horner’s syndrome.

Clavicular fractures and humerus fractures are less common fetal complications of shoulder dystocia.

Diaphragmatic paralysis would occur with injury to the phrenic nerve. However, this would present with signs and symptoms of respiratory distress including tachypnea, mild to severe respiratory distress, cyanosis, apnea, or a weak cry. Neonates with diaphragmatic paralysis may also have a brachial plexus paralysis, however given that this baby has no respiratory symptoms, this is a less likely diagnosis.
A 46-year-old man presents with sudden jerking movements of his arms. His family notes that he has become moody and irritable. His father was killed in a car crash at age 36. What pathological change is likely to be found in this man's central nervous system?

- a. Degeneration of lateral corticospinal tracts and anterior motor neurons of spinal cord
- b. Depigmentation of the substantia nigra and locus ceruleus
- c. Frontotemporal atrophy and gliosis
- d. Generalized cerebral atrophy and neurofibrillary tangles
- e. Neuronal depletion and atrophy of the caudate nuclei

**Correct answer: E**

This patient is likely suffering from Huntington's chorea, given his stereotypic arm movements and behavioral changes. The early death of his father may have masked the family history of this autosomal dominant disorder. His brain pathology would likely show neuronal depletion and atrophy of the caudate nuclei (Choice E).

Degeneration of lateral corticospinal tracts and anterior motor neurons of spinal cord (Choice A) is seen in ALS, which is characterized by motor weakness, not behavioral changes.

Depigmentation of the substantia nigra and locus ceruleus (choice B) is typical of Parkinson's disease.

Frontotemporal atrophy and gliosis (choice C) is seen in Pick's disease, a progressive dementia characterized by behavioral disinhibition.

Generalized cerebral atrophy and neurofibrillary tangles (Choice D) is seen in Alzheimer's disease.
A 27 year old man presents to the ED with severe retroorbital pain of one days duration. He says the pain is really severe and came on somewhat suddenly. The pain is primarily concentrated behind his left eye. He also noted that his eyes, especially the left, is tearing and that his nose is also dripping. He says he has had these "attacks" before. In the ED, he denies a history of trauma, neck stiffness, recent illness or URI, or vision loss. Ophthalmic exam is conducted and is found to be normal. A CT scan of the head and Lumbar puncture are normal. He is afebrile. What is the most likely diagnosis?

- a. cluster headache
- b. glaucoma
- c. migraine headache
- d. meningitis
- e. subarachnoid bleed

**Correct answer:** A

A young man with retroorbital pain associated with lacrimation and rhinorrhea is a cluster headache until proven otherwise. Often there is a history of these severe headaches. They can indeed be very painful. Glaucoma is caused by an increase in ocular pressure-which would be seen on ophthalmic exam. Also acute glaucoma is usually precipitated by some event such as watching a movie. Migraine headaches are more common in woman and do not present with lacrimation and rhinorrhea. Meningitis is important to consider in a young man with a headache but a normal lumbar puncture makes this unlikely. A subarachnoid bleed would also be caught either on a CT scan of the head or with a LP.
A 67-year-old woman comes to your office complaining of the recent onset of hallucinations. Upon questioning you discover that she was well until 4 months ago; since then she has been increasingly forgetful and irritable. Sometimes she can't seem to get her feet and legs to work, and she has fallen multiple times over the last two months. If you were to take a brain biopsy, what would you see microscopically?

- Basophilic & argentophilic, round, well-demarcated, fibrillary inclusion
- Eosinophilic intracytoplasmic inclusions
- Eosinophilic rods of paracrystalline actin arrays
- Loss of the dark neurons in the anterior midbrain
- Round, pink, dense deposit of alpha synuclein protein

**Correct answer:** E

This woman has Lewy Body Dementia, which is characterized by more rapid dementia than Alzheimer's Disease, parkinsonism (bradykinesia), hallucinations, and/or pain. You would expect to see Lewy Bodies. The answer is E -- round, pink, dense deposits of alpha synuclein protein found in the brain cortex of individuals with Lewy Body Dementia. LBD combines features of both Alzheimer's (memory problems, dementia) and Parkinson's (movement disorder).

A -- Pick bodies are basophilic and argentophilic (stain with silver), round, well-demarcated, fibrillary inclusions. They are most predominant in the frontal lobe and the hippocampi of patients affected with Frontal Temporal Dementia, also known as Pick's Disease. FTD is primarily a disorder of personality and behavior, but this patient also has motor symptoms.

B -- Negri bodies are eosinophilic intracytoplasmic inclusions found in the brains of persons infected with the rabies virus. She does not have any of the classic symptoms of fever, mania, hydrophobia, or lethargy that progresses quickly to coma and death.

C -- Hirano bodies are eosinophilic rods of paracrystalline actin arrays found in the hippocampi of individuals with Alzheimer Disease (and Creutzfeldt-Jakob Disease). The dementia of AD has a more insidious onset. Our patient was well until 4 months ago.

D -- The substantia nigra appears as dark neurons in the anterior midbrain. This part of the brain is noticeably lighter in elderly individuals with Parkinson Disease. While dementia and personality changes may be seen in PD, especially in the later stages, this is primarily a movement disorder and does not fit her symptoms.
A 13-year-old male presents to your office with a club foot, difficulty walking, and worsening scoliosis. On physical exam you hear a systolic murmur that gets softer when he lies down and louder when he stands up. He also has diminished sensation for fine touch in all extremities as well as decreased deep tendon reflexes. No one in his family has experienced anything like this. What is the cause of his condition?

a. autoimmune degeneration of oligodendrocytes  
b. an autosomal dominant trinucleotide repeat  
c. an autosomal recessive trinucleotide repeat  
d. defective or absent dystrophin  
e. defective superoxide dismutase 1

**Correct answer:** C

He has Friedrich’s Ataxia, which is due to C—an autosomal recessive trinucleotide repeat (GAA, in the infrataxin gene on chromosome 9). Huntington’s Chorea is due to B—an autosomal dominant trinucleotide repeat, and ALS is due to E—a defective superoxide dismutase 1; these are both pure motor disorders. Duchenne’s and Becker’s Muscular Dystrophies are due to D—multipabsent or defective dystrophin proteins; but the former would have killed him already, and the latter does not have sensory findings like his. Multiple Sclerosis is due to A—autoimmune degeneration of oligodendrocytes; it presents with both sensory and motor findings, as this patient does, but he’s a little young. Moreover, the usual course of MS is relapsing-remitting, which is not what is described here. The murmur describes hypertrophic cardiomyopathy.
A 28-year-old man presents with recent onset decreased urinary output, and a general malaise for the past week. Physical exam is remarkable for blood pressure of 190/94, edema, and mild flank pain. Urinalysis shows red blood cell casts. A renal biopsy is performed, with nearly all glomeruli appearing as shown.

What is the most likely pathological process, and what is the likely prognosis?

a. Goodpasture’s Disease; disease is self-limited
b. Poststreptococcal glomerulonephritis; disease is self-limited
c. Poststreptococcal glomerulonephritis; renal failure is imminent
d. Rapidly progressive glomerulonephritis; disease is self-limited
e. Rapidly progressive glomerulonephritis; renal failure is imminent

**Correct answer:** A

Rapidly progressive (crescentic) glomerulonephritis rapidly progresses to renal failure; the number of crescents roughly corresponds with prognosis. The disease may follow a flu-like syndrome, but is distinct from poststreptococcal glomerulonephritis.

Poststreptococcal glomerulonephritis classically presents with “Coca-Cola” colored urine and occurs 2 weeks after an upper respiratory infection such as strep throat. Biopsy shows a uniform “filling” of glomerular capillaries with white blood cells. Most cases fully recover.

Goodpasture’s Disease often presents similarly to rapidly progressive glomerulonephritis, but requires pulmonary involvement. This condition will also rapidly progress to renal failure if left untreated.
A 55-year-old man undergoes renal biopsy due to proteinuria and hematuria. Based on the findings shown above, on which of the following medications should this patient be started?

- a. Albumin
- b. Cisplatin
- c. Dexamethasone
- d. Epoetin alfa
- e. Labetalol

**Correct answer:** E

The biopsy shows the characteristic “onion skin” lesion of hyperplastic arteriosclerosis, indicative of a severe, relatively acute increase in blood pressure. Accordingly, the patient should be started on an antihypertensive medication such as labetalol (choice E) as soon as possible.

Albumin (choice A) can be used as a treatment in hypoproteinemic states. Although this patient presented with proteinuria, there is no evidence to suggest necessity of albumin at present.

Cisplatin (choice B) is a chemotherapeutic agent known to cause nephrotoxicity and ototoxicity. Although hematuria can be a presenting symptom of renal cancers, the biopsy shows no evidence of such a condition.

Dexamethasone (choice C) is a potent corticosteroid with mineralocorticoid and glucocorticoid effects. It would be indicated in a situation such as acute adrenal insufficiency (e.g. Addisonian crisis) to help correct hypotension with circulatory collapse. It would be contraindicated for this patient with severe hypertension.

Epoetin alfa (choice D) is a synthetic analog of erythropoietin, released by the kidney to stimulate red blood cell differentiation and proliferation in the bone marrow. It is used in the treatment of chemotherapy-related and chronic renal failure-associated anemia.
A patient comes to your office with the symptoms of renal tubular acidosis. On performing test you get a defect in the proximal tubule re-absorption of HCO3-. What type of acidosis it is characterized in

- a. Type 1
- b. Type 2
- c. Type 3
- d. Type 4
- e. Type 5

**Correct answer:** B

Type 1 leads to defect in H+/K+ ATPase of collecting tubules.

Type 2 is defect of proximal tubule HCO3- re-absorption.

Type 4 is Hypoaldosteronism
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See Explanation

A is the correct answer
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c. Poststreptococcal glomerulonephritis; renal failure is imminent
d. Rapidly progressive glomerulonephritis; disease is self-limited
e. Rapidly progressive glomerulonephritis; renal failure is imminent

correct answer: E

correct answer is E not A. It is even explained in explanation.

Thanks

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A 27-year-old woman, gravida 2, para 2, comes to the physician to have her staples removed after an elective repeat cesarean delivery. Her pregnancy course was uncomplicated. She states that she is doing well except that since the delivery she has noticed some episodes of sadness and tearfulness. She is eating and sleeping normally and has no strange thoughts or thoughts of hurting herself or others. Physical examination is within normal limits for a patient who is status post cesarean delivery. Which of the following is the most likely diagnosis?

- a. Maternity blues
- b. Postpartum depression
- c. Postpartum mania
- d. Postpartum psychosis
- e. Postpartum psychosis

correct answer: A

A. Maternity blues is the term used to describe a common postpartum reaction that occurs in 50 to 70% of postpartum patients. It is characterized by tearfulness, restlessness, and anxiety. Symptoms typically start in the first few days postpartum and resolve within 2 weeks. However, certain patients continue to have the symptoms for several weeks. Many symptoms may be seen in association with this disorder including headache, backache, fatigue, forgetfulness, insomnia, weeping, depression, anxiety, and negative feelings toward the newborn infant. Interestingly, another component of the syndrome may be episodes of elation, and such mood lability can be especially distressing for the new mother. It is unclear what the etiology of these symptoms is. Certainly, the postpartum period with a newborn can be stressful and life changing, which can certainly lead to mood changes and a number of emotional responses. Some researchers have argued that changes in hormone levels are at the root of the maternity blues, but this has never been definitively proven. This patient does not have evidence of a true postpartum depression (e.g., insomnia, lack of appetite, or anhedonia) or postpartum psychosis (e.g., bizarre thoughts) and she does not have any thoughts of hurting herself or her baby. Therefore, the most likely diagnosis is maternity blues and she should be given support and reassurance. The patient must also be cautioned, however, that if her symptoms do not resolve, or if they worsen, then she must call or return. Postpartum depression (choice B) is a depression that occurs in about 10% of postpartum women and it is more serious than the maternity blues. Symptoms may include sleep disturbances and changes in appetite. Postpartum mania (choice C) or postpartum psychosis (choice D) is a psychiatric disorder that occurs in about 1 per 1,000 deliveries. It is characterized by severe anxiety, agitation, disordered thoughts, and confusion. Hospitalization is required. Poststerilization depression (choice E) is a depression that is seen in women following a tubal ligation or other form of permanent sterilization. This patient did not have a sterilization procedure.
25 year male with a testicular mass. Investigation reveals markedly increased alfa feto protein and mildly increased HCG. What is the most likely tumor?

a. Seminoma
b. Embryonal CA
c. Teratoma
d. Sertoli cell Tumor
e. Choriocarcinoma

**correct answer:** B

Seminoma= Increased PLAP (Placental alkaline phosphatase)
Embryonal CA= Increase in AFP + mild increase in HCG
A 21 year old man has a testicular mass. On physical exam there is evidence of a testicular mass that does not transilluminate. It is firm and hard and can be palpated in the scrotum. Ultrasound examination reveals a malignancy. Serum marker for PLAP is positive. His lesion is treated and he responds well to radiation therapy. What is the most likely diagnosis?

a. seminoma
b. choriocarcinoma
c. prostatic adenocarcinoma
d. Embryonal Carcinoma
e. Yolk Sac tumor

**Correct answer:** A

This young man likely has a seminoma. There is a hard mass on exam and the PLAP (PlacentaLike Alkaline Phosphatase) is positive, this is diagnostic for seminoma. The fact that it does not transilluminate means it is not hydrocele. Also, prostate cancer is very very unlikely given this presentation and the patient’s age. Choriocarcinomas usually present with an elevated B-HcG not an elevated PLAP. These tumors are more aggressive and often are hemorrhagic. Embryonal tumors may have elevation of other markers such as both the AFP and b-HcG and do not characteristically this way. Yolk sac tumors are far less common than seminomas and usually present with an isolated elevated AFP. The default for a young man with a testicular mass is a seminoma. It is very responsive to radiotherapy.
A 65 year old woman with a family history of breast cancer had a mammogram that demonstrates a diagnosis of infiltrating ductal adenocarcinoma. She generally feels well and has not lost weight, decreased her appetite, nor is she fatigued. What is the most important prognostic indicator of her breast cancer?

- a. Age
- b. Axillary lymph node status
- c. Estrogen receptor Status
- d. HER 2 NEU receptor Status
- e. Metastatic status

**Correct answer:** E

Be careful! Do not just jump to axillary lymph node status. While it is true that this is one of the biggest prognostic factors in this woman’s breast cancer, metastatic status is more an indication of the prognosis. By definition - if there are metastatic lesions – one is more concerned about the widespread dissemination of disease to bone, liver, and brain than simply local spread to lymph nodes! If metastatic status was not here- axillary lymph node status would be the answer. Her2/neu does carry a poor prognosis- interestingly these neoplasms can be treated with the MAB traztuzamab. ER status is a good prognostic indicator because these can be treated with tamoxifen or raloxifien; meaning the neoplasm will be responsive to cutting off the hormone supply that feeds its growth.

Remember breast cancer is often caused by unopposed estrogen throughout a woman’s lifespan. Age is the biggest risk factor for breast cancer and is not a prognostic factor for disease.
A 26 year old female in her 4th month of pregnancy comes to her OB-GYN with a complaint of spotty vaginal bleeding for the past 3 days. Bleeding has been accompanied by mild nausea and vomiting over the past week. On pelvic exam, you note her uterus as being much larger than expected for her the estimated gestational age of the fetus. You perform an intravaginal ultrasound which shows a “snowstorm” appearance and no fetal heartbeat can be detected. Which of the following is most likely to be elevated in the patient's serum?

a. Estradiol
b. Lactate dehydrogenase
c. Human placental lactogen
d. Human chorionic gonadotropin
e. Alpha-fetoprotein

correct answer: D

This question stem describes a complete hydatidiform mole, a benign tumor of the chorionic villus. Molar pregnancies result from abnormal fertilization and can be of two types: partial and complete. Complete moles are the most common type in which there is a 46XX genotype (completely paternal in origin) and no embryo present. Complete moles characteristically result result in having a uterus too large for gestational age, markedly elevated hCG for gestational age, “snowstorm” appearance on ultrasound and pre-eclampsia in the first trimester. Due to the elevated hCG levels, patients will test positive on home or office based pregnancy tests.

Partial moles, on the other hand, are triploid (69XXY) and contain embryonic parts.

Estradiol (A) can be elevated in a number of ovarian stromal tumors such as thecomas and granulosa cell tumors but is normally not increased in molar pregnancies. Lactate dehydrogenase (B) is a non specific marker and can be elevated in a variety of conditions such as hemolysis or liver disease but is generally not a marker for genital tract abnormalities. Human placental lactogen (C) is normally produced during fetal development but is not abnormally elevated in the context of molar pregnancies. Alpha-fetoprotein (E) can be elevated in the context of germ cell tumors or birth defects such as Neural Tube. It is not a marker for molar pregnancy.
A 55-year-old female presents to her gynecologist complaining of discomfort in her abdomen and bloating. Pelvic ultrasound reveals bilateral adnexal masses. What is the most likely origin of the adnexal mass?

a. Metastasis from endometrium
b. Metastasis from gastrointestinal tract
c. Ovarian germ cells
d. Ovarian stroma
e. Ovarian surface epithelium

**Correct answer:** E

Over 65% of all ovarian tumors and 90% of all ovarian cancers originate from the epithelium of the ovary capsule. There are six primary types of epithelial tumors: serous, mucinous, endometrioid, clear cell, Brenner, and undifferentiated. Serous cystadenocarcinomas are the most common type of epithelial ovarian cancer, and are bilateral 65% of the time. Symptoms of ovarian cancer are often vague, and include abdominal pain, bloating, and early satiety.

Metastases from primary tumors outside of the ovary (Choice A and B), known as Krukenberg tumors, are responsible for about 5 to 10% of ovarian cancers.

Ovarian germ cell tumors (Choice C) account for 15 to 20% of all ovarian tumors. Approximately 95% of these tumors are benign. The remaining 5% that are malignant are found predominantly in children and young women, making them the most common ovarian malignancy in females under 20 years of age.

Ovarian stromal tumors (Choice D) are generally low-grade malignancies that can occur at any age.
A 55 year old women had hysterectomy and bilateral salpingectomy for uncontrolled uterine bleeding. Histological examination of the ovaries show small atretic follicles. Which of the following processes is most like cause of this histological findings?

- a. Apoptosis
- b. Metamorphosis
- c. Metaplasia
- d. Necrosis
- e. Transformation

**Correct answer:** A

A) Apoptosis is defines as programed cell death. At age 55 after menopause graffian follicles are found atretic, by the process of apoptosis.

C) Metaplasia is a intermediate stage of cancerous conversation, and hence have no role in menopause.

D) Necrosis is seen in conditions like diabetic foot, infarction.

E) Transformation is conversion of 1 type of epithelium to another, seen in Baretts oesophagus, cancerous conditions.
A 33-year-old man status-post renal transplant presents with one week of cough, fever, chills, and pleuritic chest pain. Chest x-ray is negative for consolidations. Lung biopsy is shown above.

Which of the following treatments is most appropriate for this patient?
   a. Amphotericin B
   b. Ganciclovir
   c. Oseltamivir
   d. Ribavirin
   e. Trimethoprim-sulfamethoxazole

**Correct answer:** B

This patient is suffering from atypical (interstitial) pneumonia. The biopsy shows the characteristic “owl’s eye” inclusions of cytomegalovirus (CMV) infection, a common complication of immunosuppressed organ transplant recipients. Ganciclovir (choice B) preferentially inhibits the DNA polymerase of CMV.

Amphotericin B (choice A) is a potent antifungal agent, binding to ergosterol in fungi membranes to form disrupting pores. Though patients receiving immunosuppression often have fungal infections, biopsy results in this patient reveal a different pathology.

Oseltamivir (choice C) inhibits neuraminidase of Influenza A & B to decrease release of progeny virus. It is not used in the treatment of CMV.

Ribavirin (choice D) inhibits synthesis of purine nucleotides as a competitive inhibitor of certain RNA polymerases. It is used in treatment of Hepatitis C.

Trimethoprim-sulfamethoxazole (TMP-SMX, choice E) inhibits bacterial dihydrofolate reductase and dihydropteroate synthase, and is a classic treatment for uncomplicated urinary tract infections. Though it is used in immunosuppressed patients for P. carinii prophylaxis, biopsy results show this patient has an active CMV infection that needs treatment.
A 18 yrs old college student comes to the hospital complaining of generalized body weakness and flu-like symptoms for 4 week duration. He has a temperature of 101F and coughs up yellowish sputum. The patient has taken several over-the-counter medications but has not been better. He is currently not sexually active and does not have any other chronic illness. A CXR of the patient is shown below.

**Based on this presentation, what is the most likely diagnosis?**

- a. Lobar pneumonia
- b. Rhinovirus infection
- c. Atypical pneumonia
- d. Pulmonary edema
- e. Congestive heart failure

**Correct answer: C**

Answering this question correctly requires that one understands the presentation of atypical pneumonia and the treatment indicated. In atypical pneumonia also referred to as ‘walking pneumonia’ patients do not present as sick as their X-ray look. The constellation of flu-like symptoms, cough with productive sputum, and fever for greater than 4 weeks without responding to OTC medications and a characteristic interstitial fibrosis on X-ray indicates a most likely diagnosis of community acquired pneumonia in this otherwise healthy young man.

Lobar pneumonia (choice A) is incorrect. The X-ray pattern in lobar pneumonia typically reveals consolidation of a lobe seen as a patchy infiltrate in a lobe.

Rhinoviral infection (choice B) is incorrect. Rhinovirus causes common cold which typically runs a course of about 2 weeks and subsides without treatment. Some patients might experience multiple bouts of infection with different strains of virus that will present similarly to this case, however, the X-ray pattern is not consistent with a non-complicated rhinovirus infection.

Pulmonary edema (choice D) is incorrect. Pulmonary edema usually occurs with an underlying cardiac etiology.

Congestive Heart Failure (choice E) is incorrect. The signs and symptoms and age of this patient is inconsistent with CHF. In CHF patients might experience symptoms of cough, generalized weakness, peripherally edema and/or pulmonary edema, with a decline in cardiac function.
45 yr old woman has recent onset of shortness of breath. She has a cough & 14 kg weight loss in past 6 months. A x ray shows 4 cm mass in middle lobe of right lung. Biopsy shows small cell carcinoma. Which of the following abnormality is most likely to develop in this patient?

a. Hypocalcemia  
b. Hypoglycemia  
c. Hyponatremia  
d. Polycythemia  
e. Hypotension

**correct answer**: C

Ectopic production of arginine vasopressin (AVP) is responsible for hyponatremia in case of small cell carcinoma which is a neuroendocrine tumor. 15% patient of small cell ca present with hyponatremia.
A 65-year-old man who works at Boeing factory in town presents with fatigue, weight loss, arthralgias, and a chronic, non-productive cough. On chest x-ray there is mediastinal and hilar lymphadenopathy and reticulonodular infiltrate. Which of the following restrictive lung conditions has noncaseating granulomas and a negative Kveim test?

- a. asbestosis
- b. berylliosis
- c. coal workers’ pneumoconiosis
- d. sarcoidosis
- e. silicosis

**Correct answer:** B

The correct answer is **B**—Berylliosis, a restrictive pneumoconiosis due to heavy exposure to dust or fumes containing metallic beryllium. Nuclear and aerospace industry workers are at increased risk. The chronic form of the disease presents with systemic symptoms, much like sarcoidosis, but a Kveim test is negative. On chest x-ray there are mediastinal and hilar lymphadenopathy and reticulonodular infiltrates. Berylliosis is usually insidious until very late; there is an increased risk for lung cancer.

A—Asbestosis presents with progressive dyspnea and productive cough; chest x-ray shows irregular lines. Anyone who worked in shipyards or construction before the late-1980s, or who has been exposed to old insulation from these structures, is at risk. There is an increased risk of lung cancer.

C—Coal Workers’ Pneumoconiosis is usually asymptomatic with a “tattooing effect” on chest x-ray. Risk of cancer is not increased.

D—Sarcoidosis is a systemic condition that often has a restrictive lung disease component. It may present with Mikulicz (or Sicca) Syndrome and elevated serum levels of ACE, IFN-gamma, and TNF. Enlarged “potato nodes” in the hilar area may be seen on chest x-ray, and a Kveim test would be positive. Diagnosis is generally made by exclusion.

E—Silicosis is the most common chronic occupational disease in the world; its characteristic presentation on x-ray is with “snowstorm” appearance and egg-shell calcification of hilar lymph nodes. There is increased risk of lung cancer.
A mother visits a pediatrician with her two-year-old girl because she has been scratching herself constantly. Physical exam is notable for xerosis, and skin thickening with accentuated skin lines at the neck and hips.

Which of the following statements is most correct?

a. The mechanism of this pathologic process is well understood.
b. The patient has an increased likelihood of developing asthma later in life.
c. The patient has an increased likelihood of developing lupus later in life.
d. The patient has come into contact with poison ivy.
e. This pathologic process is usually fatal.

Correct answer: B

This little girl appears to be suffering from atopic dermatitis, which often presents in the first few years of life, usually manifesting as lichenification (accentuation of normal skin lines with skin thickening). Findings often include xerosis (dry skin) and eczematous lesions. Atopic dermatitis is frequently associated with allergic rhinitis and asthma. Although patients do not always manifest each part of this “allergic triad,” the patient has an increased likelihood of developing asthma later in life (Choice B) is the best answer.

The mechanism of this pathologic process is well understood (Choice A) is incorrect. Atopic dermatitis is a disease of unknown origin, and the pathophysiology is poorly understood.

The patient has an increased likelihood of developing lupus later in life (Choice C) is incorrect. Atopic dermatitis appears to be an allergic reaction, not an autoimmune process. No association of atopic dermatitis with lupus has been demonstrated.

The patient has come into contact with poison ivy (Choice D) is incorrect. Poison ivy exposure manifests as linear vesicles on erythematous skin.

This pathologic process is usually fatal (Choice E) is incorrect. Although it is a great source of morbidity, mortality due to atopic dermatitis would be unusual.
A 77-year-old man has multiple, painful blisters located on his face, neck, and torso. The patient complains that he cannot stop scratching them. Which additional finding would suggest a diagnosis of Pemphigus Vulgaris rather than Bullous Pemphigoid?

a. Denuded skin and flaccid blisters  
b. Fever and anorexia  
c. Intact skin and tense blisters  
d. Mouth Lesions  
e. Posthealing hyperpigmentation

**Correct answer: A**

The correct answer is denuded skin and flaccid blisters. In Pemphigus Vulgaris, the lesions are intraepidermal blisters secondary to IgG autoantibodies against epithelial cell desmogleins 1 and 3. When pressure is applied to these blisters, there is separation of the epidermis (positive Nikolshy's sign), which leads to flaccid blisters and denuded skin.

Mouth lesions are always present in pemphigus vulgaris, but they also may be seen in bullous pemphigoid.

Tense blisters are found in bullous pemphigoid in which the lesions are subepidermal. However, in pemphigus vulgaris, the lesions are intraepidermal with resulting flaccid blisters. In bullous pemphigoid, the eroded skin from ruptured blisters usually reepithelializes well without expansion into the periphery as in pemphigus. New, discrete vesicles may be found at the edges of old, resolving lesions.

Posthealing hyperpigmentation may be seen with both diseases although bullous usually does not scar.

Neither disease usually involves systemic symptoms such as fever and anorexia.
A 48 year old female presents to her dermatologist after her hair dresser noticed a "scab" on her scalp that has been present for her last two appointments that bled once after she brushed over it with her comb. Patient is an avid sunbather and tells you she has used self tanning beds twice a month since she was 20 years of age. On exam, there is a raised pearly papule with an area of central ulceration and overlying telangiectatic vessels. Which of the following is most applicable to this lesion:

a. It frequently originates as a pre-existing actinic keratosis.
b. It is locally aggressive and frequently metastasizes; depth of invasion predicts risk for metastasis
c. It is a benign skin condition that requires no further workup.
d. It is locally aggressive but almost never metastasizes.
e. It is the least common form of malignant skin cancer.

correct answer: D

This stem describes the lesion of basal cell carcinoma (BCC), a malignancy that arises from the basal cell layer of the epidermis. Histologically, BCC appears as cords of basophilic staining basal cells infiltrating the underlying dermis. BCC is caused by chronic exposure to ultraviolet light and is thus most often found on the parts of the body that are chronically exposed to sunlight such as the scalp. BCC is locally aggressive and carries a very small risk of metastasis, as compared to malignant melanoma (answer choice B) which is locally aggressive and is associated with frequent metastases.

Answer A describes squamous cell carcinoma (SCC) which is often preceded by lesions of actinic (solar) keratosis. SCC are scaly nodular lesions that are often ulcerated and are often found most commonly in areas of the body commonly exposed to sunlight. Like BCC, SCC carries minimal risk for metastasis. Answer C can describe many skin lesions including a nevocellular nevus (mole) which is a benign tumor of neural crest derived melanocytes (nevus cells). Benign nevi carry no risk of malignancy and do not require further workup. Dysplastic nevi, on the other hand, can progress into malignant melanoma, and are often identified by having irregular borders, irregular coloring and/or diameter >6mm. Answer E is a distraction as BCC is the most, not least, common malignant skin tumor.
A 11 year old boy presents to clinic with his mother with the complaint of pain in the right knee. The boy states that the pain started approximately 1 month ago and often wakes him up from sleep during the night. The pain is not relieved by warm compresses but was improved by aspirin that his mother gave him during episodes a couple nights during the week prior to presentation. PE is normal without any palpable joint deformity, redness or swelling. Which of the following findings is most likely to be seen on plain films of the right knee?

a. Ill-defined mass involving the femur metaphysis with elevation of periosteum
b. Well defined 1-cm lucency surrounded by a thin rim of bony sclerosis in the proximal tibia
c. A 7-cm mass in distal femoral epiphysis with a "soap bubble" appearance
d. A 6-cm expansile mass in diaphysis of right lower femur with an "onion skin" appearance
e. A 3-cm broad based excrescence projecting from metaphysis of upper tibia

**Correct answer:** B

Question stem describes findings characteristic of osteoid osteoma, a benign bone tumor commonly found in children and young adults (age 10-20). Pain disproportionate to the tumor size and response to aspirin are classic presentations of this tumor. Most osteoid osteomas can be effectively treated by curettage and tumor removal.

(Choice A) describes findings classic for osteosarcoma: a metaphyseal location in a long bone with characteristic "sunburst" pattern from calcified osteoid and Codman’s triangle caused by the tumor lifting the periosteum. Like osteoid osteomas, these are also found in young individuals (most in males, age 10-25). Unlike osteoid osteomas, osteosarcomas are infiltrative destructive tumors that can metastasize.

(Choice C) describes findings characteristic of giant cell tumor. Giant cell tumors arise in the epiphyses of long bones mostly in patients age 20-40.

(Choice D) describes findings consistent with an Ewing sarcoma. Ewing sarcoma usually occurs in the diaphyseal regions of long bones in patients age 10-15 and are classically associated with an periosteal reaction causing the "onion skin" appearance around bone.

(Choice E) describes osteochondroma, a benign, tumor-like projection of the cartilaginous growth plate to form an exostosis (bony outgrowth). Osteochondromas are the most common benign bone tumor.
A 73y/o woman has 1cm, scaly, raised lesion on the right side of the face. A photomicrograph of tissue obtained on biopsy of the excised lesion is shown. Use which of the following is most likely to have prevented the development of this lesion.

a. Conjugated Estrogen  
b. Isoniazid  
c. Sunscreen  
d. Tretinoin cream  
e. Triamcinolone cream  

correct answer: C  
Correct answer is C. Sunscreen  
The patient has Squamous Cell Carcinoma of the skin. To prevent it, is by trying to limit exposure especially at noon. Use high-quality sunscreens, preferably with SPF (sun protection factor) ratings of at least 30.

Option A. Conjugated Estrogen is wrong because estrogens conjugated (Premarin), is commonly used for treating the symptoms of menopause including hot flashes, vaginal dryness, and vaginal atrophy.

Option B. Isoniazid is wrong because Isoniazid is the first-line antituberculosis medication in prevention and treatment.

Option D. Tretinoin cream is wrong because this cream is commonly used to improve the appearance of the skin by reducing fine lines and wrinkles, reducing roughness and improving coloration and can also be used to treat acne.

Option E. Triamcinolone cream is wrong because this cream is not used to prevent squamous cell carcinoma, it is commonly used in the post-operative period of certain cosmetic surgery procedures, notably rhinoplasty. Injected into the subcutaneous area, it may help to alleviate stubborn swelling, inflamed sebaceous cysts, and scar tissue. Other used are to treat keratosis pilaris, psoriasis, oral irritation after dental procedures. It is highly effective in the most severe cases of eczema, but may require a waiting period before subsequent treatments.
A 68 years old female presents with a uniformly brown, round lesion which appears to be "stuck on" the right side of her face. Histologically, this lesion will most likely reveal

- a. Compound nevus
- b. Dysplastic nevus
- c. Halo nevus
- d. Junctional nevus
- e. Basal cell carcinoma

**Correct answer:** A

"Stuck on" [elevated] lesion occurs Seborrheic keratosis, a benign condition which usually occurs in older individuals. Histologically, these lesions reveal hyperkeratosis with horn and pseudohorn formation. A dysplastic nevus is more likely to develop in people who have numerous nevi spread over the entire body. A halo nevus is seen keratoacanthomas.

A junctional nevus is a flat pigmented lesion.

A basal cell carcinoma appears as raised nodule with a central crater.
A woman presents to a dermatologist because she has lost almost all the hair on her body, including scalp hair, eye brows, eye lashes, arm pit and groin hair, and the fine hairs on her body and extremities. She most likely has a variant of which of the following?

a. Alopecia areata  
b. Androgenic alopecia  
c. Chronic cutaneous lupus erythematosus  
d. Lichen planopilaris  
e. Trichotillomania

**Correct answer:** A

The correct answer is A. Alopecia areata is caused by an autoimmune attack on hair follicles. It has a wide range of clinical severity, with most cases involving a localized patch of hair (which regrows within 1 year in half of the patients). The hair that does regrow may be gray or depigmented. More severe cases can involve the entire scalp (alopecia totalis) or, as in this patient, the entire body surface (alopecia universalis). These more severe cases are less likely to resolve adequately. Treatment of alopecia areata is often unsuccessful, but topical steroids are typically tried.

Androgenic alopecia (choice B) is common male pattern baldness.

Chronic cutaneous lupus erythematosus (choice C) can produce localized baldness.

Lichen planopilaris (choice D) can produce localized baldness.

Trichotillomania (choice E), also called traumatic alopecia, is alopecia due to trauma, such as hair pulling or tight braids.
A 61-year-old woman with leukemia abruptly develops an intensely itchy rash. Physical examination demonstrates multiple erythematous patches of the distal arms and legs, some of which involve the palms and soles. Some of the patches show central clearing with surrounding erythematous rings. Which of the following is the most likely diagnosis?

a. Erythema migrans chronicum  
b. Erythema multiforme  
c. Kaposi's sarcoma  
d. Psoriasis  
e. Urticaria

**Correct answer:** B

The most specific clue in the description is the presence of erythematous patches with central clearing, known clinically as "target lesions," which are associated with erythema multiforme. Both erythema multiforme and its severe, life-threatening version, known as Stevens-Johnson syndrome, are produced by immune complex deposition in dermal blood vessels. In approximately 50% of patients, no specific precipitating cause is identified. In the remainder of patients, however, a variety of causes have been implicated, including certain infections (herpes simplex, enteroviruses, Mycoplasma pneumoniae, Chlamydia, histoplasmosis), drugs (penicillin, sulfonamides, phenytoin, aspirin, corticosteroids, cimetidine, allopurinol, oral contraceptives), neoplasia (leukemia, lymphoma, multiple myeloma, internal malignancy), sarcoidosis, and foods (notably emulsifiers in margarine). Erythema migrans chronicum (choice A) also produces an annular erythematous rash with central clearing, but usually affects the thigh, groin, and axilla; it is associated with Lyme disease. Kaposi's sarcoma (choice C) causes purple lesions with no target lesions. Psoriasis (choice D) causes erythematous plaques with silvery scale but does not produce target lesions. Urticaria (choice E) causes wheals that are intensely pruritic, but does not produce target lesions.
which of following sentences would you like to ...?

a. decrease haptoglobin
b. increase haptoglobin
c. increase bilirubinemia
d. decrease direct bilirubinemia
e. increase indirect bilirubinemia

correct answer: A
An 18-year-old male has dyspnea on exertion. A wide, fixed S2 split can be heard as well as a grade 2/6 systolic murmur in the left second intercostal space. What is the likely diagnosis?

a. Atrial septal defect
b. Bicuspid aortic valve
c. Patent ductus arteriosus
d. Tetralogy of fallot
e. Transposition of the great vessels

Correct answer: A

Right-to-left shunts cause cyanosis immediately (near birth), so tetralogy of fallot (D) and transposition of the great vessels (E) can be immediately eliminated. A patent ductus arteriosus (C) also presents early and can be eliminated. A bicuspid aortic valve (B) would result in a systolic ejection murmur and paradoxical split of S2. However, an atrial septal defect can cause a fixed A2-P2 split with a murmur at the left second intercostal space (site of pulmonic valve). Although small atrial septal defects are often asymptomatic, larger ones can lead to a left-to-right shunt in the atria. The increased blood flow in the right side of the heart results in fixed wide splitting (due to increased duration of flow through the pulmonic valve, causing the valve to close later) and a systolic murmur in the second left intercostal space (because the increased flow over the valve is turbulent).
A 76-year-old obese woman with a history of type II diabetes and heart disease presents complaining of crushing sub-sternal chest pain of 30 minutes duration with pain radiating down her left arm. A blood test within 8 hours would show an increase in which of the following cardiac enzyme levels?

- a. Aspartate aminotransferase (AST)
- b. Lactate dehydrogenase (LDH)
- c. Phospholipase C
- d. Tropomyosin
- e. Troponin I

**Correct answer:** E

Of the choices listed, Troponin-I is the best enzyme level to evaluate. Troponin-I is immediately elevated for up to 8 hours post-MI and continues to stay elevated for about 7 days. Creatine phosphokinase (CPK) is also sensitive and can be used instead of Troponin-I, although this was not a choice.

AST is increased after Troponin-I and peaks later.
LDH is increased after AST and peaks later.

It is important to learn to the pathophysiology of myocardial infarctions, as well as the characteristic lab tests used to determine relative damage/timing.
A 57-year-old obese man with a history of smoking, hyperlipidemia, and hypertension presents with pain in his legs while walking. On further questioning, he reveals that the pain starts in his buttocks and extends down his thighs and into his calves. Previously, the pain disappeared with the cessation of activity, but lately it persisted in his feet even at rest. His symptoms can best be explained by which of the following?

a. Decreased permeability of endothelium  
b. Narrowing and calcification of vessels  
c. Peripheral emboli formation  
d. Thrombus formation  
e. Weakening of vessel wall

**Correct answer:** B

This patient suffers from "peripheral" atherosclerosis, a chronic inflammatory disease of blood vessels that involves the accumulation of lipids within the intima, recruitment of leukocytes and smooth muscles cells to the vessel wall, and deposition of extracellular matrix within the arterial wall. The resulting atherosclerotic plaques can cause narrowing and calcification of major arteries (Choice B). Flow-limiting atherosclerosis in limb arteries often presents as claudication. Symptoms of limb claudication are namely buttock, thigh, or calf pain precipitated by walking and relieved by rest.

Atherosclerotic lesions are thought to develop and progress in association with endothelial dysfunction, such as increased permeability (Choice A), though this is not a major contributor to symptoms of claudication. Other complications of atherosclerosis include thrombus formation with peripheral emboli formation, thrombus formation, and weakening of vessel walls. Peripheral emboli formation (Choice C) is closely associated with embolic stroke or atheroembolic renal failure. Thrombus formation (Choice D) with occlusion of vessel lumen is associated with myocardial infarction, unstable angina, and thrombotic stroke. Weakening of vessel walls (Choice E) is associated with aortic aneurysms.
29 years old patient come to the ER after car accident. He was bleeding a massive amount about 2L. How much of fluid do give him?

a. 1L  
b. 2L  
c. 3L  
d. 4L  
e. 5L

Correct answer: C
A 31 year old businessman has been experiencing chest pain with activity for the past 6 months. He says his job is stressful, but no more than usual. On exam he is noted to have a BMI of 28.5; his blood pressure is 137/83. Coronary angiogram shows 75% narrowing of the left anterior descending coronary artery. Which of the following is a major risk factor associated with this patient?

a. Obesity
b. Type A personality
c. Diabetes Mellitus
d. Lack of exercise
e. Age

correct answer: C
Choice C is the correct answer

Diabetes mellitus is a major risk factor for early, accelerated, and advanced atherosclerosis. DM is a major risk factor for macrovascular disease. In young men or premenopausal women with heart disease, DM must be considered.

Choice A, B, D, E play lesser roles in the development of atherosclerosis. The patient’s age of 30 is young for the development of heart disease. His BMI places him as overweight, not obese.
A 21 year old patient has a throbbing pain in head. He has got itching on skin and had a weight loss of 3 kg from 58 kg to 55 kg. What is the most likely underlying pathophysiology?

a. Migraine
b. Hypovolemia
c. Hypertension
d. Hypervolemia
e. None of the above

**Correct answer:** B

Due to itching and pain loss it is clearly understood that the patient is having hypovolemia. Throbbing headache indicates that the increased density of the blood is putting pressure on the brain.
A 72-year-old man has had progressive shortness of breath for the past year. On exam, rales are heard in all lung fields. Chest x-ray shows pulmonary edema and a prominent left-sided heart shadow. An echocardiogram shows that the left ventricular wall is concentrically hypertrophied. Which of the following has most likely produced these findings.

- a. Centrilobular emphysema
- b. Chronic alcoholism
- c. Silicosis
- d. Systemic hypertension
- e. Tricuspid valve regurgitation

**Correct answer:** D

Hypertension is an important cause of left ventricular hypertrophy and failure. Left-sided heart failure leads to pulmonary edema with dyspnea.

Centrilobular emphysema (Choice A) and Silicosis (Choice C) are incorrect; obstructive diseases lead to pulmonary hypertension with right-sided heart failure from cor pulmonale.

Chronic alcoholism (Choice B) is incorrect; this can lead to a dilated cardiomyopathy that affects heart function on both sides.

Tricuspid valve regurgitation (Choice E) is incorrect; right-sided heart lesions predispose to right-sided heart failure.
A 20 year old female patient arrives to your office with a chief complaint of neck swelling. She also complains of weight gain and fatigue. What is the best initial diagnostic test to order?

- Presence of antimicrosomal antibodies
- Free T4 levels
- TSH levels
- Thyroglobulin levels
- Thyroid Scan

**correct answer: C**

This patient likely has Hashimoto’s thyroiditis given her age, gender, goiter and symptoms of hypothyroidism. Although antimicrosomal antibodies would be likely be present, the initial screening test to measure thyroid function is TSH. In her case, it should be elevated because her thyroid is not producing enough thyroid hormone.
An asymptomatic 50-year-old woman has hypertension. Her urinary excretion of catecholamines is increased. A CT scan shows a suprarenal mass. Which of the following is the most likely cause?

a. Benign neoplasm of the adrenal cortex
b. Benign neoplasm of the adrenal medulla
c. Malignant neoplasm of the adrenal cortex
d. Malignant neoplasm of the adrenal medulla
e. Diffuse hyperplasia of the adrenal cortex

**Correct answer:** B

Increased urinary excretion of catecholamines suggests increased production of catecholamines, a process that occurs in the adrenal medulla.

(You should know the various layers of the adrenal glands and the respective hormones produced in each layer.)

The patient presents with symptoms of hypertension secondary to a suprarenal mass, consistent with a pheochromocytoma. You should know that such masses are much more commonly benign (only 10% malignant).

Therefore (B) benign neoplasm of the adrenal medulla is the most likely cause.

Increased production of catecholamines is consistent with
A 50-year-old woman has had a painless mass in the parotid gland for the past 8 months. A 2-cm, discrete, solid mass is found in the parotid gland on parotidectomy. Histologic examination shows a neoplastic lesion with uniform epithelial and myoepithelial cells; these cells form acini, tubules, and ducts supported by myxoid and chondroid stroma. Which of the following is the most likely complication of this type of parotid lesion?

a. Contralateral immune-mediated parotitis  
b. Hematogenous metastases to lungs and bone  
c. Ipsilateral submaxillary salivary gland neoplasm  
d. Local recurrence  
e. Regional lymph node metastases

**correct answer:** D
A 50-year-old male with a history of diabetes returns to his physician's office for review of his labs. His health maintenance labs show a hemoglobin A1c of 9.0, increased from 7.0 at his last visit. His current drug regimen includes glipizide and metformin. He reports that he takes his medications regularly, but his wife is concerned about his diabetes control. Which of the following labs would best indicate a need for insulin to control this patient's blood sugar?

a. Anti-GAD antibody
b. C-peptide
c. Fasting blood glucose
d. IGF-1
e. Random blood glucose

correct answer: B

Insulin-resistant diabetes, aka adult-onset or Type II, eventually becomes insulin dependent when overworked pancreatic beta cells stop producing insulin. One way to determine when insulin therapy is indicated is to check a C-peptide (Answer B). C-peptide is cleaved when pro-insulin is converted to insulin; normal ranges of C-peptide indicate endogenous production of insulin.

Choice A is incorrect. Anti-GAD antibody is used to diagnose Type I, or autoimmune Diabetes.

Choices C and E are incorrect. A random blood glucose >200 or a fasting blood glucose >126 is used to diagnose Diabetes. Measurement of blood glucose is not a reliable test of insulin production.

Choice D is incorrect. IGF-1 level is used to diagnose growth hormone abnormalities.
A 35-year-old woman presents to her physician complaining of headaches, increased sweating, and intermittent stabbing pain on her right side. She has no chronic medical conditions, does not take any medication, and does not smoke or use other drugs or alcohol. Her family history is significant for thyroid cancer on her maternal side, type unknown. On physical examination, the patient is pale and anxious appearing. Her blood pressure is 210/130 and her heart rate is 120. Which of the following diagnoses is most consistent with the information available thus far?

- Anxiety disorder
- Essential hypertension
- Hyperthyroid
- Multiple endocrine neoplasia type I
- Multiple endocrine neoplasia type II

**Correct answer:** E

Multiple endocrine neoplasia (MEN) type II, also known as Sipple’s syndrome, is characterized by medullary carcinoma of the thyroid, pheochromocytoma, and parathyroid tumor or adenoma. MEN type I (Choice D), also known as Wermer’s syndrome, is characterized by pancreatic, parathyroid, and pituitary tumors. Both MEN type I and II are inherited in an autosomal-dominant fashion. In this case, the clinical history includes several hyperadrenergic symptoms consistent with pheochromocytoma, including marked hypertension, tachycardia, pallor, and diaphoresis. Additional symptoms of pheochromocytoma include palpitations, anxiety, and weight loss. A common manifestation of hyperparathyroidism is hypercalcemia, which can result in kidney stones and flank pain. These findings, when coupled with a family history of thyroid cancer, should make one suspicious of MEN type II. Anxiety disorder (Choice A), essential hypertension (Choice B), and hyperthyroid (Choice C) should be included in the differential diagnosis; however, based on the patient’s presentation and lack of medical conditions or risk factors, these choices are less likely to be responsible for the patient’s complaints.
A 27-year-old female gives birth to a 8 lb, 8 oz boy but upon delivering the placenta starts to bleed heavily. The obstetrician estimates the blood loss at 2000 ml (normal <500 ml). She recovers well after IV saline infusion and 2 units of packed red blood cells. After one week, she notices that her milk never "let down" and she is unable to breast feed her child. One year later, she has still not had a menses, and she complains of fatigue, cold intolerance, weight gain despite dieting and hair loss.

What is most likely cause of these symptoms?

a. Addison's disease  
b. Cushing's disease  
c. Hashimoto's thyroiditis  
d. Pituitary adenoma  
e. Sheehan's syndrome

**correct answer: E**

The patient is suffering from Sheehan's Syndrome (option E) which is a loss of anterior pituitary function after significant blood loss suffered during delivery. During pregnancy, the anterior pituitary becomes hypervascularized and if the mother hemorrhages the pituitary can become ischemic. Without anterior pituitary function, the mother does not produce prolactin (necessary for breast feeding), does not produce FSH/LH (for ovulation), TSH (cold intolerance, weight gain, hair loss) or ACTH (loss of glucocorticoids and androgens). The treatment is to give thyroxine, glucocorticoids and possibly FSH/LH.

Addison's Disease is a bilateral loss of the adrenal glands which leads to a lack of mineralocorticoids, glucocorticoids and androgens. Usually it is thought to be caused by an autoimmune reaction.

Cushing's Disease (option B) is a ACTH secreting tumor leading to an increase in glucocorticoids and androgens (mineralocorticoids are regulated by renin-angiotensin system). Symptoms include hypertension, central obesity, buffalo hump, poor wound healing, moon facies.

Hashimoto's thyroiditis (option C) is an autoimmune cause of hypothyroidism. This would cause some of the symptoms above but would not appear so quickly after birth and would not inhibit lactation.

Pituitary Adenoma (option D) is a growth of the pituitary. It can cause Cushing's Disease if it secreting ACTH, but can also cause pituitary dysfunction by mass effect by inhibiting hypothalamic regulators from reaching the anterior pituitary. An adenoma would not present so quickly after birth.
A 41 year old woman undergoes a thyroidectomy for removal of a malignant growth. Shortly after the surgery she complains of muscle cramps and is noted to have a twitch in her cheek when the facial nerve is tapped. What is the most likely cause of her symptoms?

a. Hyperkalemia due to kidney failure
b. Hypervitaminosis D due to hyperparathyroidism
c. Hypocalcemia due to hypoparathyroidism
d. Hypocalcemia due to hypothyroidism
e. Hyponatremia due to adrenal insufficiency

correct answer: C
This patient displays muscle cramps and Chvostek's sign (tapping on the facial nerve produces a tetanic spasm) which are typical symptoms of hypocalcemia. After a thyroidectomy, the most likely cause is inadvertent removal of the parathyroid glands, leading to hypoparathyroidism (Choice C). The other choices are unlikely in this scenario.
A 36 year old male comes to your office complaining of “feeling cold” for the past 7 months. He notices that he has been using less air conditioning this summer and is always dressed in layers at work. On review of symptoms, he reports a 8 lb weight gain over the past year and says he notices some blood on the toilet paper occasionally from straining to pass his stools on the toilet. Which of the following is the most appropriate initial test to order?

a. Total triiodothyronine (T3) level
b. Total thyroxine (T4) level
c. Thyroid stimulating hormone (TSH) level
d. Radioiodine scan
e. Fine needle aspiration biopsy

correct answer: C

This stem describes a patient with a very classic clinical presentation of hypothyroidism. Characteristic symptoms include weight gain, cold intolerance and constipation, as seen in this patient. Other common manifestations include fatigue, weakness brittle hair and nails, and depression. The best screening test for suspected hypothyroidism is a TSH level. Levels of TSH indicated whether there is primary thyroid disease- and would be elevated if there is a primary thyroid disease (ie Hashimotos thyroiditis) that was the cause of this patient’s symptoms. Alternatively, levels would be low in other types of primary thyroid disease, such as Graves disease, in which there is an overproduction of thyroid hormone and thus symptoms of hyperthyroidism. Although levels of T3 and T4 (options A and B) would be expected to be low in most forms of hypothyroidism, these tests would not help indicate the underlying cause of the disease. Radioiodine scans and fine needle aspiration biopsies (options D and E) are used in the workup of thyroid nodules and masses, and would not be an appropriate first step for this patient.
A 38 year old woman complains of 4 days of vomiting. She attributes it to food allergies. She denies blood in the vomitus. She reports that her energy level “has not been the same.” Which change in concentration do you expect to see in the patient’s plasma?

a. Elevated PTH  
b. Elevated total calcium  
c. Reduced sodium  
d. Elevated potassium  
e. Elevated insulin

**Correct answer:** A

From her history of vomiting, this patient most likely has metabolic alkalosis, secondary to losses of gastric acid through vomiting. The high level of bicarbonate will be buffered by hydrogen ions from COOH groups on albumin, increasing the number of negative sites on albumin (COO\(^{-}\)). Those extra negative sites will bind calcium ions, reducing the amount of ionized calcium. Reduced levels of Ca\(^{2+}\) will lead to an increased PTH to compensate the apparently low calcium. Nonetheless the total calcium will remain normal (B is wrong).

C is incorrect. Given the dehydration from vomiting, this patient is more likely to be hypernatremic.

D is incorrect; metabolic alkalosis leads to exit of hydrogen ions from cells, in exchange for cellular penetration by potassium to maintain electroneutrality.

E is incorrect. The loss of nutrients places this patient in a catabolic state, which will results in an elevated level of glucogenic hormones such as glucagon and cortisol, not insulin.
A 13-year-old male comes to the emergency room with a one-day history of nausea and vomiting. He says that yesterday, he began having pain around his belly button and skipped dinner. Today, he felt nauseated. His mother thought he had a fever and kept him home from school. He says the pain moved to his right lower quadrant and, "it hurts there when I press it." On exam, he shows rebound tenderness and guarding in his right lower quadrant, 1/3 of the way between the right iliac crest and the umbilicus. A CBC shows an elevated white blood cell count to 14.2. What is the most appropriate next step?

a. Antibiotics to cover gram-negative organisms  
b. Appendectomy  
c. CT scan  
d. Stool guaiac exam  
e. Ultrasound of abdomen

**correct answer:** B

This patient is presenting with the classic symptoms of appendicitis: anorexia, fever, elevated white blood cell count, and periumbilical pain migrating to the right lower quadrant. In a young male with a classic story, imaging is not needed to confirm the diagnosis. It is more important to bring him to the operating room emergently to avoid the complications of a perforated appendicitis. Antibiotics are not an appropriate treatment at this time. A stool guaiac exam is unnecessary.

The step 1 and 2 questions #29 have been mixed up when viewing the answers. Please correct accordingly.
A 22-year-old Jewish female who has been experiencing right lower quadrant abdominal pain, diarrhea, and weight loss for several weeks visits the hospital. After her physician performs an unspecified procedure, a pathology report returns the displayed image (bottom image only). Which of the following is a complication of this patient’s most likely diagnosis?

a. Perforation of the appendix
b. Perianal fistulas
c. Right ovarian necrosis
d. Severe dehydration
e. Toxic megacolon

**correct answer:** B

The patient likely has Crohn's disease. The histology shows a slide typical of the transmural intestinal inflammation and fissuring ulcers found in the disease. The clinical scenario is also consistent with this diagnosis; particularly, this patient’s RLQ pain is typical as the terminal ileum is often affected. Complications of Crohn’s disease include fistulas (particularly in the perianal region, but fistulas can also form between the intestine and bladder/uterus) and abscesses. Remember that Crohn's disease can affect anywhere in the GI tract ("mouth to anus"), so aphthous ulcers can be seen in the mouth, and even the esophagus can be affected (rarely).

Toxic megacolon (E) is one of the complications of ulcerative colitis. Perforation of the appendix (A) would occur in a case of appendicitis, which would have a much more acute presentation. While malabsorption of nutrients is common in Crohn's (hence the weight loss in this patient), severe dehydration (D) would more likely occur secondary to severe diarrhea due to a bacterial infection such as E. coli or Shigella (this presentation would also be more acute). Right ovarian necrosis (C) would follow ovarian torsion.
A 67-year-old woman presents with complaints of pruritis which has developed over several months. Physical exam is notable only for yellow-tinged sclera. Laboratory values reveal an increased direct bilirubin. What is her diagnosis?

- a. Gilbert Syndrome
- b. Primary Biliary Cirrhosis
- c. Crigler-Najjar Type I
- d. Acute cholecystitis
- e. G6PD deficiency

**Correct answer: B**

Primary biliary cirrhosis (PBC) results in a direct hyperbilirubinemia through fibrosis of the biliary tree resulting in cholestasis. Anti-mitochondrial antibodies are usually present. Pruritus is usually the first symptom, jaundice develops later.

Gilbert syndrome and Crigler-Najjar types 1 and 2 would result in an indirect hyperbilirubinemia due to a defect in UDP-glucuronosyltransferase which conjugates bilirubin. Of note, Crigler-Najjar type 1 usually results in kernicterus in the neonatal period.

Glucose-6-phosphate dehydrogenase (G6PD) deficiency would also result in an indirect hyperbilirubinemia due to hemolysis.

This patient's presentation is not consistent with acute cholecystitis given time course and lack of fever or abdominal pain.
A 65 year old man undergoes a subtotal gastrectomy for gastric carcinoma. During the procedure, most of the body of the stomach was resected. There were no intra-operative complications and the immediate post-operative course was uneventful. What is the most likely long-term post-operative complication that this patient will develop?

a. Vitamin B6 deficiency  
b. Vitamin C deficiency  
c. Vitamin B1 deficiency  
d. Iron deficiency  
e. Zinc Deficiency

**Correct answer:** D

The body of the stomach is the primary location for parietal cells, and a subtotal gastrectomy that involves the majority of the body of the stomach will greatly reduce the number of parietal cells in the stomach. Parietal cells produce gastric acid and intrinsic factor. Intrinsic factor is necessary in the absorption of vitamin B12. Gastric acid has numerous functions, one of which is to aid the absorption of iron. Ingested iron comes in different forms: heme, ferrous (Fe++), and ferric (Fe+++). Ferrous iron is soluble and can be easily absorbed in the duodenum/jejunum while ferric iron is insoluble. Stomach acid is needed to decrease the pH to allow reduction of ferric iron to ferrous iron. Without adequate number parietal cells, stomach pH will not be low enough to facilitate ferric iron absorption, potentially leading to iron deficiency (choice D).

Vitamin B6 deficiency (choice A) is most commonly induced by INH and certain oral contraceptives.

Vitamin C (choice B), folate (choice C), and zinc (choice E) deficiencies are not associated with loss of parietal cells or stomach size.
A 50-year-old man with a long history of alcohol abuse and chronic hepatitis B infection present with a protuberant abdomen and scleral icterus for 3 weeks duration. The patient is in no acute distress; however, physical exam reveals a constellation of signs suggestive of liver cirrhosis and portal hypertension. Which of the following signs is a result of decreased hepatic oncotic function in cirrhotic patients?

a. Spider nevi on chest and face
b. Gynecomastia
c. Redistribution of pubic hair
d. Testicular atrophy
e. Lower limb swelling

**Correct answer:** E

Lower limb swelling (choice E) in cirrhotic patients is caused by a combination of portal hypertension and low oncotic pressure secondary to hypoalbuminemia resulting in fluid accumulating in the extracellular space. The liver is the major site of albumin production.

Spider nevi (choice A), gynecomastia (choice B), redistribution of pubic hair (choice C), and testicular atrophy (choice D) are all signs of hyperestrogenism in a man with cirrhosis. Hyperestrogenism occurs in men with cirrhosis because the liver cannot degrade estrogen and 17-ketosteriods such as androstenedione. Androstenediones are subsequently aromatized into estrogen in adipose tissue.
A 42-year-old woman presents to the ER with five hours of steadily increasing abdominal pain. The pain is associated with nausea and vomiting, and began soon after a heavy meal. Physical exam reveals an overweight middle aged woman in mild distress. She has no signs of jaundice. Her abdomen is exquisitely tender in the right upper quadrant. The remainder of the exam is normal. Ultrasound shows several 3 cm gallstones, an edematous gallbladder wall, and a positive sonographic Murphy’s sign.

For what malignancy is she at increased risk?

a. Carcinoma of the gallbladder
b. Cholangiocarcinoma
c. Hepatoblastoma
d. Hepatocellular carcinoma
e. Pancreatic cancer

Correct answer: A

This patient demonstrates the clinical picture of acute cholecystitis, which is not a risk factor for carcinoma of the gallbladder. However, multiple large stones do increase the risk of this condition. It should be noted that the vast majority of patients with cholelithiasis will not develop carcinoma of the gallbladder.

Neither cholelithiasis nor cholecystitis are associated with increased risk of cholangiocarcinoma. Risk factors include Primary sclerosing cholangitis, chronic duct inflammation, infections, and ulcerative colitis.

Hepatoblastoma is a rare malignant neoplasm of children.

Hepatocellular carcinoma risk factors include cirrhosis, Hepatitis C, and Hepatitis B.

Pancreatic cancer risk factors include smoking, a high fat diet, and chemical exposures. Of note, alcohol consumption is not a demonstrated risk factor.
A 46-year-old female with a history of painful spasms of the fingers in the cold complains of 2 weeks of fatigue, pruritus, and abdominal pain. Her exam is notable for scleral icterus, right upper quadrant pain, and a liver span of 11cm. Her laboratory values are as follows:

- AST: 110
- ALT: 102
- Alkaline Phosphatase: 256
- Total Bilirubin: 2.8
- Direct Bilirubin: 1.9

An abdominal ultrasound shows no obstruction. A liver biopsy is taken and shows destruction of small interlobar bile ducts.

What antibody test will likely be positive?

a. Anti-Scl 70
b. Anti-ds DNA
c. Anti-mitochondrial
d. Anti-smooth muscle
e. P-ANCA

**Correct answer:** C

This patient is suffering from primary biliary cirrhosis (PBC) which is most commonly diagnosed in women in their 4th and 5th decade. PBC is an autoimmune disease in which the intrahepatic bile ducts are affected. Anti-mitochondrial antibodies (option C) are commonly found. The fact that she has a history of an autoimmune disease (painful spasms of the fingers in the cold is typical for Raynaud's phenomenon) raises suspicion for this diagnosis. The elevated bilirubin and alkaline phosphatase in the absence of evidence of gall stones or obstruction of the biliary tract on ultrasound makes PBC the most likely diagnosis in this case.

Anti-Scl 70 (choice A) is seen in systemic scleroderma, which typically presents with skin tightening or esophageal dysmotility when part of the CREST variant.

Anti-ds DNA (choice B) is a specific marker for SLE.

Anti-smooth muscle antibodies (Choice D) are a marker of autoimmune hepatitis which would be expected to further elevate the AST and ALT.

P-ANCA (choice E) is often seen in primary sclerosing cholangitis (PSC), and in a variety of vasculitis diseases including Churg-Strauss. PSC is more commonly seen in patients with underlying ulcerative colitis and more commonly in men.
A 33-year-old man is seen by his gastroenterologist for progressive dysphagia over the last 3 weeks. Barium swallow shows a dilated esophagus with an area of distal stenosis, resembling a ‘bird beak’ appearance.

The treatment of this condition places the patient at a higher risk for which of the following?

a. Barrett’s esophagus
b. Esophageal squamous cell carcinoma
c. Gastric carcinoma
d. Mallory-Weiss syndrome
e. Plummer-Vinson syndrome

Correct answer: A

This patient presents with achalasia, which results from failure of relaxation of lower esophageal sphincter (LES) due to loss of the myenteric (Auerbach’s) plexus. The progressive dysphagia is caused by the stenotic LES. Treatment of achalasia includes pneumatic dilation of the LES and surgical myotomy. Because the treatment involves relaxing the LES, the patient can be more prone to acid reflux, which can lead to Barrett’s esophagus (A).

Esophageal carcinoma (B) can cause achalasia, but there is no conclusive evidence that achalasia is a premalignant lesion for esophagus cancer. In addition, the choice states squamous cell carcinoma, which tends to occur in the upper 2/3 of the esophagus and is not usually located near LES. Barrett’s esophagus, however, can eventually lead to adenocarcinoma of the esophagus.

Gastric carcinoma (C) may result from disruption of the gastric mucosa from chronic gastritis. Treatment of achalasia doesn’t influence such processes.

Treatment of achalasia doesn’t increase the risk for vomiting or retching which would cause a tear at the gastroesophageal junction (Mallory-Weiss (D). Plummer-Vinson syndrome (E) is a long-term iron deficiency anemia resulting from esophageal webs in the proximal esophagus (near the upper esophageal sphincter). Achalasia is not associated with PVS.
A 10-month-old boy presents to the emergency room with a 6 hour history of intermittent crying and refusing to feed. He had one episode of vomiting, which his parents report was green in color but free of blood. He passed loose, bloody stools on the way to the emergency room. Physical examination reveals an irritable infant with a temperature of 102.5°F and a palpable mass in the right upper quadrant. What is the most likely diagnosis?

a. Appendicitis  
b. Intussusception  
c. Meckel’s diverticulum  
d. Pyloric stenosis  
e. Volvulus

**Correct answer:** B

Intussusception is the invagination of one portion of the bowel into itself, usually the proximal portion drawn into the distal portion by peristalsis. It has a peak incidence in infants ages 5 to 12 months with a male predominance. The classic triad of intussusception includes intermittent colicky abdominal pain, bilious vomiting, and “currant jelly” stool. Neurologic signs, such as lethargy, seizure, and apnea, may also be seen and can delay diagnosis. Physical examination frequently reveals a sausage-shaped mass in the right upper quadrant. Diagnosis is with ultrasound or contrast enema, which has therapeutic as well as diagnostic value. Intussusception most commonly occurs idiopathically, but is also associated with viral infection or an anatomic lead point, such as a Meckel’s diverticulum, polyp, or lymphoma.

Appendicitis (Choice A) classically presents with pain, vomiting, low-grade fever, and anorexia. Diarrhea is an infrequent finding. A localized mass may be palpated on rectal exam, but generally not on abdominal exam.

Meckel’s diverticulum (Choice C) usually presents in the first 2 years of life with intermittent, painless rectal bleeding and intestinal obstruction. It occurs when the embryonic duct connecting the yolk sac to the intestine fails to regress completely and persists as a diverticulum connected to the ileum. Acid-secreting gastric mucosa lines the diverticulum and can produce ulcerations of the diverticulum itself or adjacent ileum, resulting in rectal bleeding. While Meckel’s diverticulum may act as a lead point for intussusception, the infant is presenting with acute symptoms of intussusception.

Pyloric stenosis (Choice D) generally presents after 3 weeks of age, but can appear as late as 5 months. It is marked by nonbilious vomiting that is typically worse after feeding. Physical exam can reveal a palpable olive-shaped mass in the mid-epigastrium, representing the thickened pylorus.

Volvulus (Choice E) occurs when a loop of bowel twists upon itself, often resulting in intestinal obstruction. It is characterized by vomiting in infancy, abdominal pain, and early satiety.
A 24-year-old woman presents with severe diffuse abdominal pain. She has a six-month history of crampy abdominal pain accompanied by intermittent bloody diarrhea. On physical exam she is found to have marked abdominal distention. Further workup reveals toxic megacolon, which requires an emergency colectomy. Pathological examination of her colon would likely reveal which of the following?

- a. cobblestone mucosa
- b. Fistulas
- c. Granulomas
- d. pseudomembrane
- e. pseudopolyps

**correct answer:** E

This patient exhibits signs and symptoms of ulcerative colitis (UC) complicated by toxic megacolon. UC is an inflammatory bowel disease that only involves the mucosa and submucosa and does not present with a pseudomembrane as in c.difficile infections (choice D) of the colon, as opposed to Crohn’s disease, which exhibits transmural inflammation.

Cobblestone (choice A), fistulas (choice B), and granulomas (choice C) are typical manifestations of Crohn’s disease, not UC.

Crohn’s disease can involve the rectum or spare it, but UC always involves the rectum.
A 45-year-old woman with a body mass index of 33 presents to her physician with a 1-day history of severe right upper quadrant pain, nausea, and vomiting. Lab studies show a serum total bilirubin level of 4.5 mg/dL. She is diagnosed with nonpigmented gallstones. Which of the following materials is the major component of these stones?

- a. Bile acids
- b. Calcium bilirubinate
- c. Cholesterol
- d. Fatty acids
- e. Phospholipids

**Correct answer:** C

The correct answer is C. This patient has a typical presentation of gallstones with right upper-quadrant abdominal pain, nausea, & vomiting. Risk factors for gallstone formation include being female, being overweight, and being over age 40. Eighty percent of gallstones are either cholesterol or mixed stones, and cholesterol comprises 70% of these mixed stones.
A 45-year-old man with a history of diabetes mellitus and gallstones comes to his doctor because of a recent 4.5-kg weight loss and fatty stools. Laboratory studies show decreased levels of pepsinogen, amylase, lipase, Insulin, and glucagon. A CT scan shows a mass in the pancreas. Based on this patient’s symptoms, which of the following substances might be secreted by this pancreatic tumor?

a. Cholecystokinin  
b. Gastrin  
c. Intrinsic factor  
d. Secretin  
e. Somatostatin

**Correct answer:** E

The correct answer is E. Somatostatin is an inhibitory hormone that is normally produced by D cells of the pancreas and inhibits gastric acid, pepsinogen, insulin, glucagon, pancreatic and small intestine fluid release, and gallbladder contraction. Somatostatin secretion is induced by amino acids, free fatty acids, glucose, glucagon, and cholinergic and beta-adrenergic neurotransmitters. It is inhibited by alpha-adrenergic neurotransmitters and insulin. Somatostatin secreting tumors commonly present with a triad of steatorrhea, diabetes mellitus, and gallstones. These tumors are most commonly found in the pancreas but can also be found in the small intestine.
One month after eating dinner at a new Indian restaurant, a man develops fatigue, malaise, and loss of appetite. He states one of his friends who also ate with him that night has been ill. On exam he has mild scleral icterus and with mild RUQ tenderness. Lab studies show AST of 70 U/L, ALT of 55 U/L, total bilirubin of 3.0 mg/dl, and direct bilirubin of 2.8 mg/dl. Which of the following test results is likely to be positive?

- a. Anti-HBs IgG
- b. Anti-HDV IgM
- c. Anti-HAV IgM
- d. Anti HBc IgG
- e. Anti-HCV IgG

**Correct answer:** C

This man has a mild, self-limiting liver disease after a meal at a restaurant. He mostly has HAV infection from contaminated food. IgM anti-HAV will indicate recent infection which will be replaced in a few weeks by IgG antibodies, giving lifelong immunity. HAV infection is short and mild with a short incubation period. Infection is the fecal-oral route.

Choice A, B, D, E are incorrect.

Hepatitis B and C requires a longer incubation period and is acquired parenterally. HDV requires coinfection with HBV.
A 45 year old man complains of having heartburn for many years, but had previously never consulted a physician. There are no findings on physical exam. An upper GI endoscopy is performed and esophageal biopsy is taken from an area of velvety mucosa above the gastroesophageal junction. Pathology shows columnar metaplasia with goblet cells. What is the most likely cause of these findings?

a. Achalasia  
b. Esophageal varices  
c. GERD  
d. Iron deficiency anemia  
e. Radiation therapy

**Correct answer:** C  
Choice C is the correct answer

This patient has Barrett esophagus which is columnar metaplasia of the lower esophageal mucosa is often a consequence of gastroesophageal reflux disease. One third of patients with Barrett’s develop esophageal cancer.

Choice A is incorrect as achalasia will show dilation of the esophagus above the lower esophageal sphincter.

Choice B is incorrect as esophageal varices result from long-standing portal hypertension.

Choice D is incorrect as iron deficiency can rarely be accompanied by upper esophageal webbing.

Choice E is incorrect as radiation therapy may produce inflammation and fibrosis but not metaplasia.
A 23 year-old male presents to the clinic for a check-up. He complains of bouts of loose bowel movements, lasting for several days at a time and occurring about 6 times in the past 6 months. His physical exam is unremarkable except for a positive fecal occult blood test on rectal exam. A colonoscopy reveals lesions in the colon and biopsy yields a diagnosis of ulcerative colitis. Several years later he gradually becomes jaundiced, with pale stools and dark urine, but no abdominal pain. The patient’s jaundice is most likely due to which of the following mechanisms?

a. Decreased production of UDP-glucuronosyltransferase and decreased ability to conjugate bile.
b. Hepatocyte destruction and increased fibrosis
c. Increased peripheral erythrocyte destruction
d. Intrahepatic and extrahepatic inflammation of bile ducts
e. Pancreatic mass obstructing the distal common bile duct

**Correct answer:** D

Ulcerative colitis is associated with primary sclerosing cholangitis (PSC) and is a result of the intra and extrahepatic destruction of bile ductules. This process is thought to be autoimmune in its mechanism, and anti smooth-muscle antibodies are found in almost half of cases but are not specific for PSC.

Choice A is incorrect; this describes Gilbert’s syndrome.

Choice B is incorrect; hepatocyte destruction does not increase in ulcerative colitis. Hepatic fibrosis is more closely associated with alcoholic cirrhosis or other causes of cirrhosis.

Choice C is incorrect; although increased peripheral destruction of erythrocytes could lead to hyperbilirubinemia and jaundice, this is not associated with ulcerative colitis. This mechanism may be more closely linked to sickle cell anemia.

Choice E is incorrect; while a pancreatic mass or tumor may obstruct the common bile duct and cause jaundice, this is not the most likely mechanism in this patient.
A 46-year-old woman with severe hypopigmentation of hands and skin, steatorrhea, osteoporosis and hepatosplenomegaly are present. Liver function test indicates increased cholesterol and anti-mitochondrial antibodies. The patient likely diagnosis is

a. Primary Biliary Cirrhosis  
b. Hepatocellular Carcinoma  
c. Wilson Disease  
d. Cholelithiasis  
e. Chronic Hepatitis

**Correct answer:** A  
More common in females above 40yrs of age. Liver Biopsy of this patient will reveal Cholestasis and anti mitochondrial antibodies confirm the test. Cholestyamine is given to for treatment.
A 49 year old male is being treated for a PE. On the 6th day of hospitalization, he is found to have a platelet count of 69000/mm³. Heparin therapy is stopped immediately to prevent which of the following complications?

a. Arterial thrombosis  
b. Heart failure  
c. Liver failure  
d. Severe bleeding  
e. Vertebral collapse

**Correct answer:** A

This patient likely has heparin-induced thrombocytopenia (HIT). Even though the platelet absolute count is lowered, HIT paradoxically causes more thrombosis than bleeding. HIT occurs when antibodies form to heparin and platelet factor IV. The antibody reacts by attaching to Fc receptors on platelets, which in turn causes platelet activation.

The typical presentation of HIT is a fall of platelet counts more than 50% in 5-10 days after initiation of heparin. The patient may present with widespread ischemic features. Discontinuation of heparin and treatment with direct thrombin inhibitors like argatroban and lepirudin is recommended.
On your rotation in the intensive care unit, you follow a patient with advanced sepsis from a central line infection. On exam you notice marked petechiae on his extremities as well as some large clots around his mouth. Concerned about disseminated intravascular coagulation, you check his labs. What would you expect?

- a. High platelets, low bleeding time, high PT, high PTT
- b. Low platelets, high bleeding time, high PT, high PTT
- c. Low platelets, high bleeding time, low PT, high PTT
- d. Low platelets, low bleeding time, high PT, high PTT
- e. Low platelets, low bleeding time, low PT, low PTT

correct answer: B

The basis of disseminated intravascular coagulation is excessive consumption of clotting factors and platelets to the point where bleeding can no longer be stopped. Thus you see a paradox of clotting and bleeding in the same clinical picture. The consumption of coagulation factors and platelets leads to prolonged PT and PTT as well as a low platelet count. Bleeding time is determined mainly by platelet count—the fewer platelets, the longer it takes for the platelet plug to be formed, the longer the bleeding time.
A 18-year-old woman presents with menorrhagia over the last several years, although her menses "have always been heavy." On further questioning, she reports having easy bruising for as long as she can remember, and that her mother also bruises easily. Which of the following is the most likely cause of her symptoms?

a. Factor V Leiden  
b. Lupus anticoagulant  
c. Hemophilia A  
d. Protein C deficiency  
e. Von Willebrand's disease

**correct answer:** E

Defects in Von Willebrand factor (E) can cause platelet abnormalities as well as a decreased amount of circulating factor VIII, and thus lead to the manifestations of 'superficial' bleeding (bruising, heavy menses, nosebleeds, etc). Von Willebrand disease is also the most common hereditary bleeding disorder.

Factor V Leiden (A) causes a hypercoagulable state. The Leiden variant of factor V cannot be inactivated by activated protein C. A deep vein thrombosis or pulmonary embolism question would be a common clinical scenario seen in a Step 1 question.

The lupus anticoagulant (B) is named such because it causes a prolongation of the PTT in vitro, but leads to hypercoaguability in vivo. It is associated with anti-phospholipid syndrome.

Hemophilia A (C) is an X-linked deficiency in factor VIII, resulting in bleeding. It is easily differentiated from Von Willebrand's disease because (1) it almost exclusively occurs in boys as it is X-linked, (2) it is more severe and will present during childhood, and (3) the bleeding is 'deep' (i.e. internally, as in joints or internal organs) as opposed to 'superficial.'

Protein C deficiency (D) is not associated with bleeding, but with hypercoagulability, due to and inability to inactivate factor V.
A 40 year old female with SLE complains of increasing fatigue over the past several months. On exam she is found to have a Hb of 9.3g/dl. A blood smear is obtained and shows normochromic normocytic erythrocytes. An iron panel is obtained with the following values:
Iron: 43
TIBC: 212
Ferritin: 368

What is likely cause for her anemia?

a. Anemia of chronic disease
b. B12 deficiency
c. Folate deficiency
d. Iron deficiency
e. Lead poisoning

**correct answer: A**

In anemia of chronic disease (option A), patients will have normochromic normocytic anemia with low iron, low TIBC and high ferritin. Anemia of chronic disease often accompanies chronic diseases like SLE, Crohn's disease, rheumatoid arthritis.

In comparison to iron deficiency anemia (option D) which is a hypochromic, microcytic anemia in which the iron is low, the TIBC is high and ferritin is low.

Folate and B12 deficiency are macrocytic in nature often with hypersegmented neutrophils.

Lead poisoning is also normocytic however, basophilic stippling will usually be seen on a smear as well.
A 26 year-old male with sickle-cell anemia presents to the emergency department with 1 day of severe epigastric and right upper quadrant abdominal pain. He also states he has shoulder and neck pain. He can recall pains like these several times over the past year, but none this severe. An abdominal ultrasound indicates several stones in the gallbladder with gallbladder wall thickening. Which of the following is true?

a. His gallstones are most likely predominantly composed of cholesterol since biliary excretion is the only way that the body excretes cholesterol
b. Chronic splenic infarcts are most likely the cause of his pain
c. His gallstones are most likely predominantly composed of bilirubin due to chronic hemolysis
d. He has gallstone pancreatitis
e. He has acute chest syndrome caused by microvascular pulmonary infarcts

**correct answer:** C

The answer is C. Chronic extravascular hemolysis occurs in sickle cell disease, resulting in hyperbilirubinemia which may result in formation of gallstones, namely bilirubin (or pigment) stones. These gallstones are commonly the cause of cholecystitis and gallstone pancreatitis. Gallbladder pain may refer to the shoulder or neck due to irritation of the diaphragm which is innervated at the levels C3, C4 and C5. Cholecystectomy would be indicated in this patient.

Choice A is incorrect; Although mixed stones (with primarily a cholesterol component) are the most common type of stones, one would expect this patient to have bilirubin stones due to the chronic state of extravascular hemolysis (due to his sickle cell anemia).

Choice B is incorrect; although chronic splenic infarcts are common in young patients with sickle cell, this is unlikely the cause of his current symptoms which include right upper quadrant abdominal pain.

Choice D is incorrect; there is no evidence of gallstone pancreatitis. Gallstones are the most common cause of pancreatitis worldwide. A stone passes through the cystic and common bile ducts and may occlude the area of the ampulla of Vater, causing bile reflux into the pancreas. This results in non-infectious inflammation of the pancreas, manifested by elevated levels of serum amylase and lipase, which are not mentioned in the above case.

Choice E is incorrect; while acute chest syndrome is common in patients with sickle cell anemia, this patient most likely has acute cholecystitis. Acute chest syndrome results from microvascular infarcts of the pulmonary parenchyma due to sickled erythrocytes occluding the intravascular lumen. The most common cause of death in patients with sickle cell anemia is lung disease due to repeated, lifelong damage.
The graph shown depicts the oxygen-Hb dissociation curve. The curve in red represents a normal physiologic condition; the grey dashed curve represents behavior in Condition X. Of the choices below, what condition best explains the findings?

- a. Alkalosis
- b. Anemia
- c. Carbon monoxide (CO) poisoning
- d. Increased 2,3-BPG
- e. Polycythemia

**correct answer:** C

In Condition X, the curve is shifted to the left, indicating facilitated loading of oxygen onto hemoglobin, and corresponding decrease of oxygen unloading in tissues. (In other words, P50, the partial pressure of oxygen at 50% hemoglobin saturation, is decreased; although oxygen content is graphed here, the relationship to hemoglobin saturation is inherent.) Additionally, the curve is shifted downward, indicating overall oxygen content is decreased. Several factors may shift the curve to the left, and several factors may shift it downward, but only one of the above does both: Carbon monoxide (CO) poisoning (Choice C) is the correct answer.

Alkalosis (Choice A), whether of metabolic or respiratory origin, shifts the curve to the left. However, it does not affect the overall oxygen content of blood, and therefore cannot explain the downward shift of the curve. Of note, decreased temperature, decreased 2,3-BPG, and CO poisoning will also shift the curve to the left.

Anemia (Choice B) decreases the overall oxygen carrying capacity of the blood, which could explain the downward shift of the oxygen content curve. However, anemia does not affect hemoglobin affinity for oxygen, and therefore cannot explain the leftward shift of the curve.

Increased 2,3-BPG (Choice D) decreases the affinity of hemoglobin for oxygen, resulting in a curve shift to the right, not left as seen here. Though a rightward shift results in an increased P50, the phenomenon facilitates unloading of oxygen in tissues; it is also seen with increased temperature, decreased pH (acidosis), and increased 2,3-BPG.

Polycythemia (Choice E) increases the overall oxygen carrying capacity of the blood, which could result in an upward, not downward, shift of the oxygen content curve. Additionally, polycythemia does not affect hemoglobin affinity for oxygen, and therefore cannot explain the leftward shift of the curve.

Image adapted from: http://commons.wikimedia.org/wiki/File:Hb_saturation_curve_unlabeled.png
A 26 year old male recently returned to the United States after a 3 month stay in Africa where he was diagnosed with Plasmodium vivax infection. After 4 days of treatment with primaquine, he developed fever, chills, back pain and dark urine. A CBC was drawn which revealed a hemoglobin of 6.7g/dL, WBC count of 12,000/mm³, and a platelet count of 362,000/mm³ and a reticulocyte count of 10% Peripheral smear reveals RBCs that are dark in color and somewhat deformed. A urine dipstick done in your office is positive for blood. Which of the following tests is most likely to identify the etiology of this patient's pathology?

a. Mean corpuscular hemoglobin concentration
b. Direct Coomb's test
c. Heinz body preparation
d. Hemoglobin electrophoresis
e. Serum ferritin concentration

correct answer: C

This stem describes a classic manifestation of glucose-6-phosphate dehydrogenase (G6PD) deficiency which was provoked by the recent exposure to primaquine. Oxidant stress (induced by medications such as primaquine, dapsone, sulfa, fava beans or infection) leads to an acute hemolytic anemia. Since these patients are G6PD deficient, they are unable to neutralize the hydrogen peroxide that is a by-product of the RBC metabolism. The accumulation of the hydrogen peroxide damages the RBC membrane and denatures the hemoglobin, forming precipitants known as Heinz bodies which can be identified on a peripheral smear. The confirmatory test for G6PD deficiency is the Heinz body preparation (diagnostic during periods of active hemolysis) which would be followed up with an enzyme assay for G6PD as a confirmatory study.

Choice A, mean corpuscular hemoglobin concentration (MCHC) - or the average hemoglobin in RBCs -- would be normal in G6PD deficiency. MCHC is most notably increased in hereditary spherocytosis and decreased in microcytic anemias. Direct Coomb's test (choice B) is used to detect IgG or complement on RBC membranes, and is used to diagnosed autoimmune hemolytic anemias. G6PD deficiency is not immune mediated and thus a direct Coomb's test should be normal in this patient. Hemoglobin electrophoresis (choice D) is used to diagnose hemoglobinopathies such as sickle cell disease and thalassemias and would be normal in pure G6PD deficiency. The onset of symptoms following exposure to a certain medication help distinguish G6PD deficiency from a hemoglobinopathy in which baseline abnormalities would be expected (regardless of medication exposure or infection). Serum ferritin (choice E) would be abnormal in iron deficiency anemia or iron overload disorders, but should also be normal in this patient.
A young female patient presents with a history of epistaxis. A increased bleeding time is confirmed. A Ristocetin assay shows no agglutination of the patient's platelets. There is no agglutination with the the addition of ristocetin and vWF either. Which bleeding disorder does the patient have?

a. Bernard-Soulier disease
b. Glanzmann's Thrombasthenia
c. Idiopathic Thrombocytopenic Purpura
d. Thrombotic Thrombocytopenic Purpura
e. von Willebrand's disease

**Correct answer:** A

Ristocetin will activate vWF which can then bind GpIb to promote platelet agglutination. In von Willebrand's disease, addition of vWF to a Ristocetin assay will fix the endogenous vWF deficit and lead to platelet agglutination.

In Bernard-Soulier disease, GpIb is lacking so addition of vWF to a Ristocetin assay will demonstrate no platelet agglutination.

In Glanzmann's Thrombasthenia, agglutination in response to Ristocetin will be normal. However, agglutination in response to ADP will not occur.
What is the reason for the dark morning void seen in Paroxysmal Nocturnal Hemoglobinuria?

a. Respiratory acidosis that occurs while sleeping
b. Respiratory alkalosis that occurs while sleeping
c. Overnight concentrating effect of the urine
d. Circadian rhythm
e. Metabolic Acidosis

Correct answer: C

It was originally thought that respiratory acidosis that occurs at night while sleeping was behind the dark morning void. However, hemolysis occurs throughout the day so the reason for the dark morning void is the concentration of urine that occurs overnight while sleeping.
Which of the following has the worst prognosis?

- a. ALL
- b. AML
- c. CLL
- d. CML
- e. APL

**Correct answer:** B

AML - 5 year survival is 21%
CML - 89%
ALL - 87%
CLL - 75%
APL - Treatable with ATRA
You are a doctor treating patients at a Mount Everest base camp at an elevation of 10,000 feet. What type of polycythemia do you expect to see in most of the patients?

a. Relative Polycythemia
b. Absolute Polycythemia (appropriate)
c. Absolute Polycythemia (inappropriate)
d. Polycythemia Vera
e. Polycythemia Vera Type 2

**Correct answer: B**

Relative Polycythemia Vera - Occurs due to a loss of plasma volume which will hemoconcentrate the blood. Polycythemia is RELATIVE to the now decreased plasma volume.

Absolute Polycythemia (appropriate) - Hypoxia leads to EPO release which increases RBC production. This increase in RBC production is appropriate because it is counter-acting the hypoxia. The high altitude at the Mount Everest base camp would be an appropriate stimulus. (Correct Answer)

Absolute Polycythemia (inappropriate) - EPO release in the absence of hypoxia.

Polycythemia Vera - Clonal expansion of myeloid cells independent of EPO.

Polycythemia Vera Type 2 - Does not exist
What derangement is seen in Relative Polycythemia?

- a. Decreased plasma volume
- b. Increased RBC mass
- c. Increased EPO
- d. Decreased EPO
- e. Elevated SaO2

**Correct answer:** A

Relative Polycythemia is due to plasma volume loss which hemoconcentrates the blood. RBC mass, EPO, and SaO2 will be normal.
A patient with Polycythemia Vera complains of pruritus after a hot shower. What is the cellular etiology of the complaint?

a. Basophils  
b. Neutrophils  
c. Eosinophils  
d. Mast cells  
e. Platelets

correct answer: D

The increase in temperature from the warm shower will cause mast cells to degranulate.
A 26 year old Asian female comes to the ED after being hit by a car. She complains of severe abdominal pain and distention. An emergency laparotomy shows a bleeding splenic laceration, and a splenectomy is performed. There are no post-operative complications. She has no significant past medical history. She drinks alcohol occasionally but denies cigarettes or illicit drugs. She is a mechanical engineer. Which of the following vaccines is recommended?

a. Hepatitis A  
b. Hepatitis B  
c. Meningococcal  
d. Pertussis  
e. Salmonella  

Correct answer: C  
Following splenectomy, patients are at increased risk for severe infection by encapsulated organisms like S. pneumoniae, n. meningitidis and H. influenzae. S. pneumoniae is the most common cause of sepsis in such patients. Patients undergoing splenectomy should receive vaccinations against these organisms, ideally 2 weeks before surgery or, if splenectomy was emergent, at least 2 weeks after. Nevertheless, most physicians prefer to vaccinate the patient during their hospital stay in order to avoid losing the patients to follow-up.
A 68 year old man with metastatic lung cancer is brought to the emergency room by his daughter, who reports that her father has been increasingly confused and lethargic over the past week. He denies any recent head trauma or any other medical problems. He reports taking chemotherapy but no other medications. On physical examination, his vital signs are stable, no jugular venous distension is present, and cardiac and lung exams are unremarkable. A CT scan does not show any brain metastases.

Laboratory results show:
Sodium: 124 mEq/L
Potassium: 4.6 mEq/L
Chloride: 102 mEq/L
Bicarbonate: 22 mEq/L

What is the most appropriate next step in managing this patient?
- a. Administer diuretics and water orally
- b. Administer isotonic saline through a peripheral IV
- c. Administer water orally
- d. Restrict all oral intake
- e. Restrict intake of water

**correct answer:** E

This patient likely has SIADH, as a result of his lung cancer. He is hyponatremic, but euvolemic, as indicated by a normal jugular venous pressure and clear lungs. Other causes of euvolemic hyponatremia could be defects in glucocorticoid synthesis/release or hypothyroidism. In this patient, however, excess ADH secretion is likely, leading to the retention of free water and producing hyponatremia. The first step in management of SIADH is to restrict the intake of water, producing a gradual rise in serum sodium.
A 55-year-old female presents to her physician with mild fatigue. Past medical history is unremarkable. She is taking no medication. No abnormalities are detected on physical examination. The only abnormality detected on routine blood testing is an elevated calcium [2.96 mmol/L (11.9 mg/dL)] and a serum inorganic phosphorus of 0.65 mmol/L (2 mg/dL). An immunoreactive parathyroid hormone level is undetectable. The most likely etiology for this patient’s high serum calcium is

a. primary hyperparathyroidism
b. malignancy
c. hypervitaminosis
d. hyperthyroidism
e. familial hypocalciuric hypercalcemia

correct answer: B

Patients who present with hypercalcemia and hypophosphatemia should be thought of as having an excess of parathyroid hormone activity. Patients with nonparathyroid hormone–like mediated hypercalcemia, such as those with excessive levels of vitamin D caused by intoxication or sarcoidosis or by increased bone turnover as in hyperthyroidism, would not be expected to have a low serum phosphate. Patients with familial hypocalciuric hypercalcemia, an autosomal dominant trait, often have normal or slightly low levels of immunoreactive parathyroid hormone. Thus, those with hypercalcemia and hypophosphatemia without elevated levels of parathyroid hormone are likely to have the hypercalcemia of malignancy. The clinical setting usually but not invariably makes this diagnosis obvious. It is clearly recognized that many solid tumors, including carcinomas of the lung and kidney, may produce a parathyroid hormone–related protein that will not be identified by the currently available assays that detect true parathyroid hormone elaborated from the parathyroid gland. This parathyroid-related protein synthesized by tumors bears striking amino acid homology to that of native parathyroid hormone with regard to amino acids 1 through 13 but is thereafter unique. In fact, it is recognized that the majority of patients with cancer and hypercalcemia have humoral hypercalcemia, as determined by elevated urinary cyclic AMP excretion.
A 33-year-old female presents with fatigue and malaise of 2 months duration. On questioning, the patient has also admits some tingling in her feet. Laboratory results indicate that this patient has a macrocytic, megaloblastic anemia. The physician thinks the patient may have a folate deficiency, and chooses to treat her with intravenous folate alone. Which of the following is the most likely consequence of this treatment decision?

- Induction of Corneal Neovascularization
- Masked signs of neural damage
- Night Blindness
- Pellagra
- Premature Alopecia

**Correct answer:** B

The two most common causes of megaloblastic anemia are vitamin deficiencies, especially folate and vitamin B12 (cobalmin). Folate deficiency is common in nutritionally deprived people and occasionally in pregnant women. B12 deficiency is classically associated with pernicious anemia, though may also result from a shortened bowel or Crohn's disease (B12 is uptaken in the ileum).

This patient likely has pernicious anemia and a B12 deficiency. While folate is indicated in such patients, intravenous vitamin B12 is required to correct underlying neurological problems. B12 deficiency can produce neurological disturbances such as optic neuropathy, subacute combined neurodegeneration, parasthesias and abnormal myelination. While administration of folate alone would moderately correct the anemia, it would mask underlying neurological pathologies.

Clinically, anyone with megaloblastic anemia suspected to be caused by a B-vitamin deficiency should receive both B12 and folate to prevent this problem.

The other answers are detractors and result from alternative vitamin deficiencies.

Corneal Neovascularization (A) can be observed with vitamin B2 (riboflavin) deficiency.

Night Blindness (C) is a consequence of Vitamin A (retinol) deficiency.

Pellagra (D) refers to the classic triad of Dermatitis, Dementia and Diarrhea, caused by vitamin B3 (niacin) deficiency.

Premature Alopecia (E) often results from Vitamin B5 (pantothenate) deficiency.
A 55-year-old man with an 80 pack year history of smoking and 3 year history of diabetes complains of bloodshot eyes, drooping of his left eyelid, a constricted left pupil and inability to sweat on his face. He has experienced some hoarseness in his voice and worsening of his cough over the previous 3 months. Which of the following is the most likely explanation of his symptoms?

- a. Lambert-Eaton Syndrome
- b. Lobar Pneumonia
- c. Malignant mesothelioma
- d. Pancoast’s Tumor
- e. Trauma to the chest

**Correct answer: D**

This patient presents with the classic triad of Horner’s Syndrome (ptosis, miosis and anhidrosis), caused by compression of the cervical sympathetic plexus. Pancoast’s Tumor, a carcinoma that occurs at the apex of the lung, commonly presents with Horner’s Syndrome in affected patients.

Lambert-Eaton Syndrome (A) is an autoimmune disorder than is often accompanied by lung carcinomas, specifically oat cell CA. Autoimmune antibodies created by the host immune system that are normally directed towards the tumor are thought to accidentally target presynaptic calcium channels at the neuromuscular junction, causing symptoms similar to myasthenia gravis.

Lobar pneumonia (B) would not cause Horner’s Syndrome, but would produce an intra-alveolar exudate and consolidation. This patient’s history of smoking should suggest a more serious lung disorder such as carcinoma.

Malignant Mesothelioma (C) is most often caused by inhalation of small particles such as asbestos or silica and involves inflammation of the mesothelium (the pleura, peritoneum or pericardium). The patients will most often have a history of industrial exposure to these toxins. Remember that smoking does not increase the risk of this lung cancer.

Trauma to the chest (E) is the least likely cause of Horner’s Syndrome, in the clinical context given. Although physical damage to the sympathetic plexus may result in unilateral symptoms as given here, they would likely present with more global symptoms, and be indicated in the history.
The parents of a two-year old boy mention to his pediatrician during a well-visit that over the past six weeks he has had increasing difficulty walking and maintaining his balance. On further questioning, the parents relate that their son always seems to be sick. On physical exam, the pediatrician notes dilated blood vessels in the eyes and the patient’s parents state that he neither has problems sleeping nor any seasonal allergies. The serum levels of which substances would most help establish the diagnosis?

- a. AFP (alpha-fetoprotein)
- b. IgA
- c. Glucocerebrosidase + Sphingomyelinase
- d. CEA (carcinoembryonic antigen)
- e. ADA (Adenosine Deaminase)

**Correct answer: A**

The patient has symptoms of ataxia-telangiectasia syndrome: the difficulty maintaining his balance evidences ataxia and in this syndrome, telangiectasia appears in the eyes or the skin. Since IgA, IgG, and IgE levels are decreased, the child is also at risk for an increased frequency of infections. Immunoglobulin levels are depressed because this syndrome results from a deficiency of a tumor suppressor/serine-threonine kinase that links double-stranded breaks in DNA to cell-cycle arrest. Since lymphoid cells have rapid turnover, they will have a high rate of DNA processing errors and so will be more severely affected by a defect in DNA mismatch repair than other cell types with a lower rate of turnover. Alpha-fetoprotein levels are elevated in ataxia-telangiectasia syndrome.

Answer choice B is wrong because although IgA levels are decreased in ataxia-telangiectasia syndrome, this is a very nonspecific test. (Remember that to rule-out a diagnosis, one should use a test with a very high specificity to minimize false-positives).

Glucocerebrosidase is deficient in Gaucher’s disease and Sphingomyelinase is deficient in Niemann-Pick’s disease types A and B. While deficiencies in one or both of these enzymes often results in ataxia, they rarely cause telangiectasias and also present with other systemic complications such as hepatosplenomegaly, leukopenia, and dyslipidemias. These two disease are included on the differential for ataxia-telangiectasia syndrome because in some cases they have a similar early presentation. However, the presence of ocular telangiectasias rules-out these diseases. Furthermore, neither of these diseases is a sequela of ataxia-telangiectasia syndrome.

CEA is a tumor marker for ovarian cancer. Ovarian cancer is not a common sequela of ataxia-telangiectasia syndrome.

Answer choice E is wrong because although the child has recurrent infections, there is no mention of the occurrence of severe debilitating infections such as those seen in SCID (Sever Combined Immune Deficiency), a form of which occurs when adenosine deaminase, an enzyme that converts adenosine to inosine is deficient. Also, ADA is not known to be decreased in ataxia-telangiectasia syndrome.
A 84 year-old female arrives at the emergency department complaining of fever for past 4 days and painful bumps that appeared on her hand this morning. On physical exam, the patient has a temperature of 39 C, and a new regurgitant murmur. Inspection of the finger nails reveals microsplinter hemorrhages as well as the findings in the photo above.

Blood cultures are sent that grow Strep bovis.

What is the most common malignancy associated with this presentation?

a. Breast cancer  
b. Colon cancer  
c. Gastric cancer  
d. Lung cancer  
e. Thyroid cancer  

**correct answer:** B  
This patient is suffering from bacterial infective endocarditis. She satisfies the Duke Criteria (new murmur, and Strep bovis, janeway lesion in the photo, osler nodes in the photo, and splinter hemorrhages). Step bovis endocarditis is associated with colon cancer, most often due to translocation of the bacteria across a disrupted colon/mucosa barrier from the malignancy (option B).

None of the other malignancies listed is associated with an increased risk of endocarditis.
An 24-year-old man is evaluated for new onset hemoptysis. He felt well until 3 days ago, when he developed an upper respiratory tract infection with cough, nasal congestion, and rhinorrhea. On physical examination was unremarkable except for a heart rate of 104/min and blood pressure of 170/100 mm Hg.

Laboratory findings
- Hemoglobin: 9.5 g/dL
- Leukocyte count: 12,500/μL
- Platelet count: 200,000/μL
- Blood urea nitrogen: 67 mg/dL
- Creatinine: 3.8 mg/dL
- Complement (C3): 135 mg/dL
- Complement (C4): 40 mg/dL
- Antinuclear antibodies: Negative
- Urinalysis: + dysmorphic RBC and RBC cast

Chest radiograph reveals bilateral fluffy pulmonary infiltrates.

Which of the following is the most likely diagnosis?

a. Systemic lupus erythematosus
b. Postinfectious glomerulonephritis
c. Goodpasture’s syndrome
d. Henoch-Schönlein purpura
e. Wegener’s granulomatosis

**Correct answer:** C

Goodpasture’s syndrome or anti-glomerular basement membrane disease is a rare condition characterised by rapid destruction of the kidneys and of the lungs. Goodpasture’s syndrome is an autoimmune disease produced due to IgG antibodies produced against the basement membrane causing damage via a type II hypersensitivity reaction. Patients usually present with upper respiratory tract symptoms and hemoptysis as seen in this case. Immunoflorescence studies staining for IgG deposits typically reveals the a linear pattern. There is often an increase in C3 and C4 complement proteins.

Systemic lupus erythematosus (choice A) is incorrect. Lupus nephritis is least likely in this patient. SLE is more common in females than in males. Furthermore, lupus nephritis usually arises within 5 years of diagnosis. The question describes a new onset hemoptysis and URI symptoms which are typically not associated with lupus nephritis.

Postinfectious glomerulonephritis (choice B) is incorrect. post-streptococcal glomerulonephritis is a disorder of the glomeruli (glomerulonephritis), or small blood vessels in the kidneys, following a streptococcal infection. The key to answering this question correctly is to know that in PSGN the URI occurs 2 weeks or more before the onset of renal symptoms. Our patient above has concurrent URI, hemoptysis with acute glomerulonephritis thus making PSGN less likely.

Henoch-Schönlein purpura (choice D) is incorrect. HSP is a systemic vasculitis characterized by deposition of immune complexes containing the antibody IgA in the skin and kidney. It occurs mainly in young children. Typical symptoms include palpable purpura (small hemorrhages in skin), joint pains and abdominal pain.

Wegener’s granulomatosis (choice E) is incorrect. Wegener’s granulomatosis is a form of vasculitis that affects the lungs, kidneys and other organs. Wegener’s granulomatosis is part of a larger group of vasculitic syndromes, all of which feature the presence of an abnormal type of circulating antibody termed ANCA (antineutrophil cytoplasmic antibodies) and affect small and medium-size blood vessels. This category includes Churg-Strauss syndrome and microscopic polyangiitis. In order to answer question correctly one must recall that in most cases of Wegener's granulomatosis ANCA is positive, yet the patient had an negative ANCA serology, making this diagnosis less likely. Patients often have a characteristic saddle-nose deformity and perforated nasal septum and other systemic manifestations including arthritis, hearing impairment, conjunctivitis, scleritis and sensory neuropathy.
A 28 year old male who have been recently treated for *P. aeruginosa* and *S. aureus* infections now complains of wheezing, failure to thrive and recurrent pain that radiates to the back. Chest X ray shows airway obstruction. Genotype of the gene shows a numerous deletion of three nucleotides that codes for phenylalanine. This patient is likely to develop?

a. Mental Retardation  
b. Tuberculosis  
c. Mousy Odor  
d. Azoospermia  
e. Ashma

**Correct answer:** D  
This patient is suffering from cystic fibrosis. The classic signs are bronchiectasis, pancreatic insufficiency, male infertility and hepatic cirrhosis. Choice D is the correct answer because the Azoospermia and infertility are found in 95% of males who survive cystic fibrosis to adulthood.
While examining the breathing motion of your 35 year old male patient, you notice ribs 6-8 move freely during inhalation but do not appear to have equal recoil during exhalation. What would be the osteopathic diagnosis?

a. Exhalation dysfunction ribs 6-8
b. Inhalation dysfunction of Rib 1
c. Inhalation dysfunction ribs 6-8
d. T 6 - 8 NReSr
e. Inhalation dysfunction of rib 5

**Correct answer:** C

In osteopathic diagnosis, the freedom is what is used for naming lesions. Ribs 6 - 8 move freely into inhalation but are restricted in exhalation therefore it is an exhalation restriction of ribs 6 - 8 and an inhalation dysfunction.
A 65-year-old woman reports to her primary care physician with a complaint of right knee pain that has been progressively worsening for the past 2 years. An x-ray of the knee shows joint space narrowing, osteophytes, and sclerotic changes. These X-ray findings are characteristic of which of the following?

- a. Normal Aging
- b. Degenerative Joint Disease
- c. Osteopetrosis
- d. Paget's Disease
- e. Rheumatoid Arthritis

**Correct answer:** B

Joint space narrowing, osteophytes, and sclerosis are characteristic of Degenerative Joint Disease (DJD, previously known as Osteoarthritis). Osteophytes are unlikely in rheumatoid arthritis. Paget's disease would show a diffuse demineralization of the bone that would probably result in a pathological fracture.
A 45 year-old female presents to your office complaining of itchy eyes for the last two months. She has no other past medical history. As an addition to her chief complaint, she also requests information about vaginal lubricant because sexual intercourse with her husband has been increasing painful due to vaginal dryness for the last four months. Her vitals are as follows: blood pressure 110/70, heart rate 70 bpm, temperature 98.8°F, respiratory rate 16. On head and neck exam, she has significant ocular dryness and conjunctival injection, cracking of her lips and parotid enlargement. Her cardiac, pulmonary and abdominal exam are unremarkable.

Which of the following is this patient most at risk for because of her medical diagnosis?

a. B cell lymphoma
b. Cushing's syndrome
c. Pheochromocytoma
d. Syringomyelia
e. T cell lymphoma

correct answer: A

This patient has Sicca Syndrome (dry eyes, dry mouth, vaginal dryness, no arthritis) and is most at risk for B cell lymphoma compared to the other choices. Sicca syndrome and Sjogren's syndrome are very similar except Sicca presents without the arthritis that is characteristic of Sjogren's. Sjogren's classically affects females between 40 and 60 years of age. B cell lymphoma of the salivary glands due to parotid enlargement is the most common neoplastic disorder related to Sjogren's.

Cushing's syndrome is due to an increase in cortisol because of a variety of causes, but not because of Sjogren's.

Pheochromocytoma is the most common surgically correctable cause of hypertension. It is usually associated with neurofibromatosis and multiple endocrine neoplasias.

Syringomyelia is an enlargement of the spinal cord that leads to pressure atrophy and hydrocephalus. It commonly presents in patients with Arnold-Chiari malformation.

T cell lymphoma is not increased due to Sjogren's, and is commonly seen with mediastinal masses in children.
A 49-year-old man presents with recurrent episodes of acute pain and swelling of his left big toe. He is a lawyer and notes that these episodes usually occur after entertaining clients with elaborate dinners. On physical examination, his left big toe is erythematous, edematous, and exceedingly tender to palpation. Analysis of synovial fluid from the joint reveals needle-shaped crystals that appear yellow when viewed with parallel light. Which of the following is the most likely etiology of his condition?

a. Autoimmune disease  
b. Calcium pyrophosphate dehydrate crystal deposition  
c. Mechanical injury  
d. Monosodium urate monohydrate crystal deposition  
e. Neisseria gonorrhoeae infection

**Correct answer:** D

The clinical picture described above is consistent with gout, a metabolic cause of arthritis in which monosodium urate crystals are deposited in joints and other tissues secondary to hyperuricemia. A number of proteins (including IgG, complement, & fibronectin) bind to these crystals and activate inflammatory cascades. Neutrophils phagocytose crystals via Toll-like receptors.

This inflammatory response is responsible for the acute and painful episodes of arthritis and bursitis, which often occur at night and generally subside over the course of 3 to 10 days. Acute gout is usually monoarticular and most commonly affects the metatarsophalangeal joint of the great toe.

The clinical picture described is most consistent with gout, which is not an autoimmune disease (Choice A).

Calcium pyrophosphate dehydrate crystal deposition (Choice B) in cartilage is responsible for pseudogout, a condition that clinically resembles gout. The calcium pyrophosphate crystals display positive intracellular positive birefringence, in contrast to the positive intracellular negative birefringence of monosodium urate crystals.

Osteoarthritis (Choice C) is the result of mechanical injury. It does not present as the clinical picture described above.

Neisseria gonorrhoeae infection (Choice E) is the most common form of bacterial arthritis. It is generally monoarticular and most commonly affects the knee. Risk factors are the same as for sexually transmitted infections.
A four year old boy presents for a checkup and sits on the floor. When asked by his pediatrician to stand up, he uses his hands as illustrated in the above diagram. What is the pathophysiology of his likely condition?

- a. Autoantibodies against the acetylcholine receptor at the neuromuscular junction
- b. Autoantibodies against voltage-gated calcium channels at the neuromuscular junction
- c. Deletion of the dystrophin gene
- d. Demyelinating plaques along peripheral and central nerves
- e. Toxin that inhibits acetylcholine release at the neuromuscular junction

**Correct answer:** C

This patient is exhibiting Gower's sign, a classic manifestation of Duchenne’s Muscular dystrophy, which is caused by a deletion or mutation of the dystrophin gene (choice C).

The other choices are causes of muscle weakness but are unlikely in this patient.

- Autoantibodies against the acetylcholine receptor at the neuromuscular junction (choice A) are typical of myasthenia gravis.
- Autoantibodies against voltage-gated calcium channels at the neuromuscular junction (choice B) are seen in Lambert-Eaton syndrome, a rare paraneoplastic autoimmune disease.
- Demyelinating plaques along peripheral and central nerves (choice D) are seen in multiple sclerosis.
- Toxin that inhibits acetylcholine release at the neuromuscular junction (choice E) refers to botulinum toxin.
A 62-year-old woman comes to your office complaining of a swollen and painful right knee. You aspirate the joint, and in the fluid are trapezoid-shaped, positively-birefringent crystals. Which of the following is not associated with her rheumatological condition?

a. Hemochromatosis  
b. Hyperparathyroidism  
c. Hyperuricemia  
d. Hypomagnesemia  
e. Hypophosphatemia

**correct answer:** C  
The crystals in the joint fluid are calcium pyrophosphate dihydrate crystals. She has pseudogout, also known as chondrocalcinosis. It is associated with hemochromatosis (A), hyperparathyroidism (B), hypomagnesemia (D), hypoposphatemia (E), and hypothyroidism. It is not associated with hyperuricemia (C—gout) or hypercalcemia.
A 45-year-old woman presents with a two-week history of recurrent left-sided chest pain occasionally radiating to her left arm. She notes this has happened intermittently for the past five years, but is somewhat worse over the past six months. She has no significant medical history and takes no medication aside from a multivitamin. She denies alcohol and cigarette use. Physical exam reveals a healthy appearing woman of normal weight, in no apparent distress. Auscultation of the heart and lungs is unremarkable. An office EKG is normal; it was without the ST changes noted on EKG two weeks ago during her first episode of chest pain. What is the most appropriate pharmacologic treatment?

a. Aspirin  
b. Enalapril  
c. Metoprolol  
d. Nifedipine  
e. Propranolol

**Correct answer: D**

This patient is suffering from variant (Prinzmetal's) angina, characterized by atypical chest pain in an otherwise healthy individuals without significant risk factors for coronary artery disease. Its pathophysiology is thought to be related to coronary artery vasospasm resulting from autonomic dysregulation and possibly endothelial dysfunction. This is similar to other conditions characterized by vasospasm, including Raynaud's syndrome. Like Raynaud's, variant angina is particularly amenable to calcium channel blockers.

Aspirin (choice A) should be used with caution and preferably avoided in individuals with variant angina because it decreases production of prostacyclin. (Per UpToDate.)

ACE inhibitors such as enalapril (choice B) are a reasonable choice for patients with CHF or hypertension in the setting of diabetes mellitus, but is not first-line therapy for variant angina. Similar for metoprolol (choice C).

Nonspecific beta-blockers like propranolol (choice E) are contraindicated because they can actually exacerbate variant angina. This is possibly due to interference with the autonomic dysfunction that underlies the condition.
Chronic cocaine consumption diminishes one's reward pathway. Repeated use becomes necessary to maintain reward, thus leading to addiction. Cocaine increases synaptic concentrations of which of the following neurotransmitters?

- a. Acetylcholine and Neurotensin
- b. Adenosine and GABA
- c. Norepinephrine and Dopamine
- d. Serotonin and Histamine
- e. Vasopressin and Somatostatin

**Correct answer:** C

Cocaine binds to the monoamine uptake transporter and blocks reuptake at the synaptic cleft of monoamines--dopamine, norepinephrine, and serotonin. The higher concentration of DOPAMINE SPECIFICALLY, results in downregulation of dopaminergic receptors at the post-synaptic membrane. The decreased dopaminergic signaling during normal physiological conditions is what leads to diminished reward and addiction.
An 18-year-old girl from Southeast Asia presents with symmetric ascending weakness below her knees. She has been unable to walk for 3 days. Physical exam reveals mild hypotension and papilledema. Upon respiratory support and administration of IV immune globulins and plasmapheresis, her condition resolves after 2 weeks. Which of the following is the most likely etiologic agent of her condition?

a. Acid sphingomyelinase deficiency
b. Arylsulfatase A deficiency
c. Infection with C. jejuni
d. Infection with JC virus
e. Microsatellite instability on chromosome 4

**Correct answer:** C

The girl presents with classic Guillain-Barre syndrome, involving inflammation and demyelination of peripheral nerves and motor fibers of ventral roots (causing motor disturbances). The symmetric muscle weakness begins in the distal extremities. In some cases, there is autonomic dysfunction involving hyper or hypotension and cardiac arrhythmias. Guillain-Barre is associated with infections such Herpes Family Viruses (especially CMV or EBV) or Campylobacter jejuni that result in immune attack of peripheral myelin. Elevated CSF protein results in papilledema.

Acid sphingomyelinase deficiency (A) causes Niemann-Pick disease, a lysosomal storage disorder resulting in organomegaly and neuromuscular disturbances. These patients are young children who do not live past adolescence.

Arylsulfatase A deficiency (B) may cause metachromatic leukodystrophy (ML), a lysosomal storage disorder. Patients with ML experience central and peripheral demyelination causing ataxia and dementia.

Infection with JC virus (D) in AIDS patients may cause a demyelinating disorder known as progressive multifocal leukoencephalopathy.

Microsatellite instability (E) on chromosome 4 results in Huntington’s Disease and involves neurodegeneration of the basal ganglia. This presents much later in life and cannot be reversed.
You are volunteering at a primary care clinic and seeing a new patient. He tells you he has been recently diagnosed with a brain tumor, but cannot remember where the tumor is located. You cannot locate his radiographic studies. He tells you that he has been suffering from seizures over the past month, as well as smell and hearing hallucinations. Based on your knowledge of neurophysiology, these symptoms are most likely caused by a tumor in which area of the brain?

a. Brain Stem  
b. Cerebellum  
c. Frontal Lobe  
d. Parietal Lobe  
e. Temporal Lobe

Correct answer: E

The location of a brain tumor typically affects its presentation. Temporal lobe tumors may lead to seizures and hallucinations that can be olfactory or auditory. If located on the dominant temporal lobe, the patient may also develop Wernicke's aphasia.

Frontal lobe lesions that overly the motor cortex can lead to paresis. Occipital lobe lesions may lead to visual problems or hallucinations. Brain stem tumors may lead to upper and lower motor neuron signs. Cerebellar tumors, more common in children, may lead to dysarthria and signs of increased intracranial pressure.
A mother brings to the clinic her 8 years old girl, because she is very short, and has a mild deviation of vertebral column, the doctor also noticed cafe au lait spots which are also present in her father, replied her mother, through slit lamp examination by an ophthalmologist identified three pigmented iris hamartomas, the doctor concluded that she has a autosomal dominant disease that can be associated with:

a. hemangioblastoma of retina/cerebellum/medulla
b. cardiac rhabdomyomas
c. cortical and retinal hamartomas
d. increased vanillylmandelic acid levels in urine
e. defect of fibroblast growth factor receptor 3

Correct answer: D

This patient has von Recklinghausen disease or neurofibromatosis type 1, a autosomal dominant disease where findings are: cafe au lait spots, neural tumors, lisch nodules (pigmented iris hamartomas). Also skeletal disorders (scoliosis), optic pathway gliomas, pheochromocytoma, On long arm of chromosome 17. Increased in VMA levels urinary says that this patient has also pheochromocytoma.

A) more often associated with von Hippel Lindau disease, tumor suppressor gene VHL 3p.
B) cardiac rhabdomyomas and C) cortical and retinal hamartomas are both found in tuberous sclerosis with seizures, mental retardation, renal cysts and renal angiomyolipomas.
E) defect of fibroblast growth factor (FGF) receptor 3, found in achondroplasia, autosomal dominant cell signaling defect of (FGF) receptor 3. Results in dwarfism; short limbs, but head and trunk are normal size. Associated with advanced paternal age, normal reproductive life.
An 83-year-old male comes to the clinic with his wife, who says he has been increasingly forgetful. Yesterday he went to the store for milk and didn’t come back for 3 hours. A few months ago, he forgot the names of his grandchildren. He has not experienced any motor difficulties or sudden changes in personality. Which of the following is true about the underlying pathophysiology of his neurological condition?

- b. Neurofibrillary tangles are intracellular aggregates of macrotubules.
- c. Amyloid plaques are correlated to dementia severity.
- d. Neurofibrillary tangles are hyperphosphorylated tau glycoproteins.
- e. Amyloid plaques are amorphous aggregates of amyloid beta-peptide sheets.

**Correct answer:** E

This patient has Alzheimer Disease. The correct answer is E: Amyloid plaques are amorphous aggregates of amyloid beta-peptide sheets. They are extracellular and are not correlated to dementia severity (C). Neurofibrillary tangles are extracellular aggregates of macrotubules and hyperphosphorylated tau protein (not beta amyloid, A). They are not intracellular (B). Tau is a neurofilament, not a glycoprotein (D).
A 32-year-old man recently recovered from an *Campylobacter jejuni* infection and is now complaining of symmetrical weakness in the distal lower extremities. Which cell type has been damaged?

- a. Motor Neurons
- b. Lateral Corticospinal Tract
- c. Schwann Cells
- d. Muscle Fibers
- e. Ependymal Cells

**Correct answer:** C

The patient is suffering from Guillain-Barre syndrome which can occur after an infection with *Campylobacter jejuni*. After the infection, there can be a type 2 hypersensitivity reaction against the myelin of schwann cells.
A histological section of the spinal cord at C3 shows a lesion in the right medial posterior column. What deficit did the patient have?

- a. Loss of vibration sense on the right side, T6 and above
- b. Loss of vibration sense on the right side, T7 and below
- c. Loss of vibration sense on the left side, T6 and above
- d. Loss of vibration sense on the left side, T7 and below
- e. Loss of vibration sense on the left side, entire body

(correct answer: B)

The dorsal columns travel in the posterior portion of the spinal cord. They are responsible for functions such as 2-point discrimination, vibration sensation, and tactile sensation.

The medial portion of the tract is known as the fasciculus gracilis and is responsible for sensation for T7 and below.

The lateral portion of the tract is known as the fasciculus cuneatus and is responsible for sensation between T6 and C1.

Lesions in these fasciculi will produce ipsilateral lesions because there has not yet been decussation. The dorsal columns decussate in the medulla. (Contrast this with the spinothalamic tract which decussates at the level of the sensory receptor)
On a routine physical exam, a 25 year old woman admits to drinking “a lot of water.” Upon questioning, she says she drinks the equivalent of about 15 L of water a day; she has to urinate about every hour. She says she has felt thirsty ever since a car accident three years ago when “I lost a lot of blood and hit my head.” A routine blood chemistry shows a serum sodium of 144. You decide to perform a water deprivation test. Which of the following lab results after the test would suggest the patient does not have central diabetes insipidus?

- a. Low levels of ADH
- b. Elevated levels of ADH
- c. Low urine osmolarity
- d. Elevated plasma osmolarity
- e. Hypernatremia

**Correct answer:** B

Patients with central diabetes insipidus (DI) have functionally low levels of antidiuretic hormone (ADH). In order to maintain a normal plasma osmolarity, they must increase their intake of free water. The other condition that can present similarly is primary polydipsia, which is a psychiatric "addiction to water." A water deprivation test can differentiate between the two conditions.

Patients with central DI have no ADH release from the posterior pituitary despite physiological stimuli. Thus after a test they will have abnormally low levels of ADH, low urine osmolarity, elevated plasma osmolarity, and hypernatremia.

A patient with primary polydipsia will have increased levels of ADH, increased urine osmolarity, normal plasma osmolarity, and normal sodium levels (ADH helps to hold onto free water).
A 42 year old black woman presents to clinic complaining of swelling around both her eyes. The patient also adds that she recently has been only urinating twice a day which is much less than her usual. Further questioning reveals a few months history of subjective fevers, malaise, and non-focal arthralgias. Vital signs show a low-grade fever and a blood pressure of 155/90. Urine analysis reveals moderate proteinuria (3g/day) and RBC casts. Blood tests reveal a hemoglobin of 9.6g/dL, BUN of 29mg/dL, and a creatinine of 2mg/dL. What is the most likely cause of this patient’s symptoms?

a. Glomerulonephritis caused by anti-GBM antibodies
b. Congestive Heart Failure caused by uncontrolled hypertension
c. Glomerulonephritis caused by immunocomplex deposition
d. Glomerulonephritis caused by T-cell production of cytokines
e. Nephrotic syndrome caused by immunocomplex deposition

correct answer: C

This patient’s symptoms and lab findings suggest a nephritic syndrome secondary to systemic lupus erythematosus (SLE). Diffuse proliferative glomerulonephritis is the common subtype of glomerular disease in SLE. Nephritic syndrome can be identified by hypertension, edema (periorbital), oliguria, hematuria, RBC casts, proteinuria (>150mg/day but <3.5g/day). The mechanism for GN in this patient is mediated by immunocomplexes circulating in the blood (i.e. DNA--anti-DNA) and depositing in the glomeruli causing the activation of the complement system which results in neutrophil recruitment and subsequent glomeruli damage (choice C).

GN caused by anti-GBM antibodies (choice A) is the mechanism seen in Goodpasture syndrome.

CHF (choice B) is unlikely in this patient without signs of pulmonary or lower extremity edema. This clinical scenario is more likely to be a GN than CHF.

GN caused by cytokines (choice D) is unlikely because cytokines, in the setting of glomerular disease, cause the GBM to lose its negative charge resulting in more of a nephrotic syndrome rather than nephritic (i.e. minimal change disease).

Nephrotic syndrome (choice E) is characterized by a more severe proteinuria (>3.5g/day), hypoalbuminemia (<3.5mg/dL), generalized pitting edema, and hypercholesterolemia. Although certain SLE-glomerular disease can have nephrotic characteristics caused by immunocomplex deposition, this patient has GN demonstrated by mild proteinuria, HTN, and RBC casts.
A 33 year old male returns to his doctor for follow-up after a routine examination reveals a serum creatinine concentration of 2.8mg/dl. Urinalysis shows 3+ protein as well as dysmorphic red blood cells, numerous white blood cells, and several red cell casts. Renal biopsy reveals crescentic changes. The above image shows the immunofluorescence under microscopy. What other clinical findings would you most likely find in this patient?

a. Cough and hemoptysis
b. Malaise, fever, weight loss, and arthralgias
c. Malar rash
d. Recent skin infection
e. Sinus tenderness

Correct answer: A

The image is consistent with linear immunofluorescence in a patient with crescentic glomerulonephritis. In the setting of GN, linear immunofluorescence is virtually pathognomonic for anti-GBM antibody disease or Goodpasture syndrome. Goodpasture syndrome is an extension of anti-GBM antibody disease defined by pulmonary involvement including shortness of breath, cough, and hemoptysis (choice A). Pulmonary involvement, usually consisting of alveolar hemorrhage, can affect up to 70% of patients.

Sinus tenderness (choice E) in the setting of GN suggests a possible diagnosis of Wegener’s granulomatosis. Wegener’s can also have crescentic glomerular changes; however, it is considered a pauci-immune GN with little to no immunofluorescence under microscopy.

Malar rash (choice C) in the setting of GN suggests a possible diagnosis of systemic lupus erythematosus. SLE can also have crescentic glomerular changes; however there would be a granular pattern on immunofluorescence.

Constitutional symptoms (choice B) in the setting of GN suggest a vasculitic diagnosis (i.e. Wegener’s granulomatosis, SLE, microscopic polyarteritis), none of which have a linear pattern on immunofluorescence.

A recent skin infection (choice D) in the setting of GN suggests a possible diagnosis of poststreptococcal GN. Poststreptococcal GN can also have crescentic glomerular changes; however there would be a granular pattern on immunofluorescence.
A 15-year old female presents to her pediatrician complaining of blood in her urine. She is otherwise healthy, with the exception of a sore throat approximately 2 weeks before. Review of systems is notable only for swollen ankles, which she denies having experienced before. On physical exam, her blood pressure is 145/85 and she has 1+ lower leg edema bilaterally. What serum study is most likely to be elevated?

a. Amyloid  
b. Anti-glomerular basement membrane  
c. Anti-streptolysin O  
d. Complement  
e. Immunoglobulin A

**Correct answer:** C

The most common cause of nephritic syndrome is poststreptococcal glomerulonephritis. It develops approximately 10 to 14 days after a group A beta-hemolytic streptococcal infection, most commonly of the upper respiratory tract but also of the skin. Features of poststreptococcal glomerulonephritis include hematuria, edema, and hypertension. Serum levels of anti-streptolysin O may be elevated. The condition is generally self-limited and resolves in a matter of weeks to months. Rare cases may develop into rapidly progressive glomerulonephritis.

Elevated levels of amyloid (Choice A) are not found in poststreptococcal glomerulonephritis. Acute phase serum amyloid A proteins may be elevated in several chronic inflammatory diseases including amyloidosis, atherosclerosis, and rheumatoid arthritis.

Anti-glomerular basement membrane (Choice B) is found in Goodpasture’s syndrome, a form of rapidly progressive glomerulonephritis.

Complement (Choice D) levels are usually decreased in poststreptococcal glomerulonephritis.

Immunoglobulin A nephropathy, or Berger’s disease, is the most common cause of glomerulonephritis worldwide. Asymptomatic, recurrent hematuria may occur 24~48hrs (not weeks) following an upper respiratory infection or exercise. IgA levels may be elevated.
A mother rushes her 4 year old son into the emergency department after finding him passed out in the garage. She says that she found him unresponsive, lying face down on the floor of their garage, and when she tried to revive him by giving mouth to mouth, she noted a sweet taste on his lips.

On physical exam, the patient is unarousable with temperature of 37.5 degrees, BP 110/60, HR 95 and RR of 20. The rest of the exam is unremarkable.

A stat metabolic panel reveals:
Na: 135
K: 3.4
Cl: 99
HCO3: 16
BUN: 18
Cr: 0.9
Glucose: 99

A urine microscopy reveals envelope-shaped crystals. What was the most likely cause of this child’s coma?

- Alcohol
- Aspirin
- Diabetic ketoacidosis
- Ethylene glycol
- Methanol

**Correct answer:** D

This child is suffering from an anion-gap acidosis (135-[16+99]=20). The mnemonic MUDPILES is useful for remembering the causes of anion-gap acidosis (Methanol, Uremia, DKA, Paraldehyde, Iron/INH, Lactic acid, Ethylene glycol, Salicylates). In this case, the child is found in the garage and has envelope shape crystals on urine microscopy, which are found in Ethylene glycol ingestion which is found in anti-freeze (option D). The crystals are formed with calcium-oxalate which precipitates due to the ethylene glycol

All the other choices could potentially anion-gap acidosis but would not cause these types of crystals in the urine nor the sweet odor on the lips. While a sweet odor could be smelled in diabetic ketoacidosis, this would be the etiology in someone with a history of diabetes or a less acute prodrome.
A 35 year-old male with no past medical history arrives at the emergency department complaining of dark urine. The patient says that this is the first time that he has ever had this problem, and he denies any kidney disease. On further questioning, the patient admits to starting a new intense weight lifting program. This morning he spent 4 hours lifting heavy weights. On physical exam, there is noted muscle weakness in all extremities. His urinalysis shows:
- blood++
- protein ++
- glucose neg
- leukocyte esterase neg
- nitrites neg

Urine microscopy is negative for RBCs and shows some epithelial cells.

Blood tests are notable for CK 1000 U/L

What is most likely cause of this patient's dark urine?
- a. Creatine kinase (CPK)
- b. E. coli
- c. Haptoglobin
- d. Myoglobin
- e. Red blood cells

Correct answer: D

This patient is suffering from rhabomyolysis due to his intense exercise program that is leading to a breaking down of the muscle. This condition is diagnosed by muscle tenderness on exam and a high CK. The muscle breakdown leads to a leak of myoglobin into the blood stream that is excreted in the kidney. The myoglobin (option D) causes the urine to turn dark and is reactive with the dip stick in urinalysis. However, urine microscopy does not show any RBCs and therefore they are not the cause of the color change (option E). Myoglobinuria can cause renal failure and this patient needs to be given intravenous fluids to protect the kidney.

Options A, B, and C will not cause the urine to change color and are not associated with a urinalysis positive for blood.
A 26-year old G1P0 woman with no past medical history presents at 18-weeks gestation for a routine check up. On physical exam she is hypertensive and has bilateral edema. Urinalysis reveals 3+ proteinuria. What is her most likely diagnosis?

- a. Eclampsia
- b. Intrauterine growth retardation
- c. Polyhydramnios
- d. Hydatidiform Mole
- e. Gestational diabetes

**Correct answer:** D

This patient's hypertension, non-dependent edema, and proteinuria during pregnancy are characteristic of preeclampsia. Preeclampsia before 20-weeks gestation is highly suggestive of a hydatidiform mole. Only seizures would qualify this patient for a diagnosis of eclampsia (A).

Intrauterine growth retardation (IUGR) and polyhydramnios are diagnoses based on ultrasound examination (B and C).

No evidence of elevated glucose or reduce glucose tolerance is provided in the question (E).
A 16 year old female comes to your clinic and says she has not started her periods yet. She looks like a normal, well-adjusted female. On physical exam, you note that her external genitalia appears female, but her vagina ends in a blind pouch and there is no cervix. Ultrasound examination reveals no uterus, but what appears to be intrapelvic gonads. Karyotype analysis reveals that the patient is 46,XY. What is the most likely diagnosis?

- a. Androgen Insensitivity Syndrome
- b. Female pseudohermaphroditism
- c. Intersexuality
- d. True hermaphroditism
- e. Turner's Syndrome

correct answer: A

The patient has a male karyotype and testes, but unambiguous female external genitalia. This is characteristic of Androgen Insensitivity Syndrome (A), in which androgen receptors of the labioscrotal folds and urogenital folds are deficient or defective. Therefore, external genitalia develops as if there were no androgens. Because there is no ambiguity in the external genitalia, this rules out intersex, female pseudohermaphroditism, or true hermaphroditism.
A 28-year-old pregnant mother with no significant past medical history brings her 3-year-old daughter to the pediatrician because her daughter has had a 5 day history of a rash and high fever. On physical exam, the temperature of the child is 38.6 degrees Celsius and the infant has an erythematous rash covering her cheeks sparing the forehead and mouth.

This infection most likely places the fetus the mother is carrying at risk for what condition?

a. Adenovirus
b. Aplastic crisis
c. Chickenpox
d. Hydrops fetalis
e. Influenza

**Correct answer:** D

The child is most likely suffering from an infection with parvovirus B19, the cause of erythema infectiosum, which is also known as "Fifth disease" or "slapped cheek disease" due to its distinctive rash on the face. The disease presents with a high fever and rash that is often described as "lacy." It usually covers the face looking like a "slapped cheek" but may cover the body as well. For immune-competent people, the infection is not dangerous and there are no longterm sequelae; however, the disease in pregnant women has been associated with hydrops fetalis, a severe form of anemia in the fetus. Hydrops fetalis is usually fatal for the fetus, although intrauterine blood transfusions may help.

In patients with sickle cell or hereditary spherocytosis, parvovirus may cause an aplastic crisis (option B); however, the mother does not have a significant past medical history.

Adenovirus (option A) and influenza (option E) can cause fevers in children and adults; however, they do not usually cause rashes and they will be associated with other symptoms like running nose (rhinorrhea), joint pain, headache.

Chickenpox (option C) is caused by the varicella zoster virus, a member of the herpes family. VZV can cause a fever and a rash, and can infect both children and adults; however, the rash is usually all over the body and is described as blister-like lesions in different stages of healing that resemble "dew drops on a rose petal."
A 17-year-old girl is brought to the pediatrician by her mother who is concerned that she has not yet started to menstruate. On physical exam, you notice that the girl is overweight, suffers from acne, and has facial hair. Which disease is the girl most at risk to develop?

- Alopecia
- Cervical cancer
- Diabetes
- Hypertension
- Ovarian cancer

correct answer: C

This girl may be suffering from polycystic ovarian syndrome (PCOS), a constellation of anovulation, amenorrhea, hirsutism, and obesity. This syndrome is characterized by a hyperandrogenic state, in which chronic anovulation leads to elevated levels of estrogen and androgens. Many patients develop insulin resistance and hyperinsulinemia, and there is an increased incidence of type II diabetes mellitus in these patients.

Hirsutism, not alopecia (Choice A), is common in PCOS. While obese patients are at risk for hypertension (Choice D) and other cardiovascular diseases, the spectrum of PCOS includes increased insulin resistance and hyperinsulinemia, putting them most at risk for diabetes. Patients with PCOS are at increased risk of endometrial hyperplasia and cancer due to the overaccumulation of uterine lining in response to unopposed estrogen. These patients are not at increased risk of cervical (Choice B) or ovarian (Choice E) cancer. Women who are not currently interested in fertility often are put on cyclic progestins or Dep-Provera to decrease this risk.
A G1P0 female at 15 weeks presents to the emergency department with vaginal bleeding for 2 days. In addition to bright red blood spotting her underwear, she also complains of some abdominal cramping. She denies passing blood clots or tissue. On exam, clotted blood can be seen in the vaginal vault and the cervix is closed. Which of the following is the most likely diagnosis?

a. Complete abortion
b. Incomplete abortion
c. Inevitable abortion
d. Missed abortion
e. Threatened abortion

**Correct answer:** E

A threatened abortion is any intrauterine bleeding before 20 weeks without cervical dilation and no passage of fetal tissue by history or physical exam. A spontaneous abortion, also known as a miscarriage, is a pregnancy that ends before 20 weeks. The type of spontaneous abortion is defined by if and how much of the products of conception (POC) have passed and whether the cervix is dilated.

A complete abortion (Choice A) is defined by the complete expulsion of all POC. On exam, the cervix is closed.

An incomplete abortion (Choice B) is defined by partial expulsion of some, but not all, POC. The cervical os is dilated to allow for the further passage of fetal tissue.

An inevitable abortion (Choice C) is defined by vaginal bleeding and an open cervical os. There has been no passage of POC but the cervical dilation makes a viable pregnancy unlikely.

A missed abortion (Choice D) is defined by the death of the fetus with complete retention of POC.
A 22-year-old female presents to her OBGYN with complaints of an increase in vaginal discharge. She describes the discharge as thin and yellow with a strong "fishy" odor. She has been in a monogamous relationship for the last several years. Which of the following findings would you most likely expect on exam?

a. KOH prep with branching hyphae and spores  
b. Negative whiff test  
c. Strawberry cervix  
d. Wet prep with clue cells  
e. Wet prep with flagellated protozoa

**correct answer:** D

The clinical picture described is classic for bacterial vaginosis (BV). Bacterial vaginosis develops when there is disruption in the normal balance of bacterial flora in the vagina. Although BV is thought to be polymicrobial, Gardnerella vaginalis is commonly present in cultures. Many patients with BV are asymptomatic; however, those that are symptomatic often complain of a profuse, nonirritating discharge that is characteristically thin, yellow, and carries a fishy amine odor. Diagnosis can be made with a wet prep of a vaginal swab that reveals clue cells, or vaginal epithelial cells that are diffusely covered with bacteria. A positive whiff test (Choice B), performed by adding KOH to a vaginal prep, is considered to be pathognomonic of bacterial vaginosis.

Cervical erythematous punctate epithelial papillae, or “strawberry” cervix (Choice C), is characteristic of infection with Trichomonas vaginalis. This unicellular, anaerobic flagellated protozoan is transmitted sexually and gives rise to a yellow, gray, or green discharge that may be frothy in appearance and gives off an unpleasant odor. Diagnosis is made via microscopic examination of wet preps of vaginal swabs in which the flagellated protozoa are visualized (Choice E).

Infection with Candida albicans can give rise to symptoms of pruritis, burning, dysura, dyspareunia, and "cottage cheese" discharge. Diagnosis is usually made by microscopic examination of a KOH prep of the vaginal discharge in which branching hyphae and spores are visualized (Choice A).
A 27-year-old female presents to the emergency department complaining of vaginal bleeding and right-sided abdominal pain. Physical examination is significant for an adnexal mass palpable on the right. Urine pregnancy test is positive. Transvaginal ultrasound is negative for an intrauterine pregnancy. Which of the following is a risk factor for this patient’s condition?

- a. Bacterial vaginosis infection
- b. Monogamous relationship
- c. Oral contraceptive pill use
- d. Prior intrauterine pregnancy
- e. Prior tubal surgery

**Correct answer:** E

The clinical scenario is that of ectopic pregnancy, in which the zygote implants outside of the uterine cavity. Patients who present with vaginal bleeding and abdominal pain should be evaluated for ectopic pregnancy as a ruptured ectopic pregnancy can rapidly lead to hemorrhage, shock, and death. Risk factors that predispose patients to extrauterine implantation include history of sexually transmitted infections or pelvic inflammatory disease, multiple sexual partners, prior ectopic pregnancy, and prior tubal surgery.

Bacterial vaginosis infection (Choice A), monogamous relationship (Choice B), oral contraceptive pill use (Choice C), and prior intrauterine pregnancy (Choice D) are not known risk factors for ectopic pregnancy.
After a mastectomy with axillary node dissection for breast cancer 1 year ago, a 55 year old woman has experienced swelling in her left arm. Exam shows firm skin over the left arm with soft underlying tissue. The arm is not painful or discolored. About 2 months she developed a skin infection. Which of the following best describes these findings?

a. Lymphedema
b. Subclavian arterial thrombosis
c. Thrombophlebitis
d. Tumor embolization
e. Vasculitis

**Correct answer: A**

A mastectomy with axillary node dissection leads to disruption and obstruction of lymphatics in the axilla. This obstruction of lymphatics gives rise to lymphedema, a condition that can be complicated by cellulitis.

Subclavian arterial thrombosis (choice B) is incorrect as thrombosis can lead to a cold, blue, painful extremity.

Thrombophlebitis (choice C) is incorrect as this is a complication from venous stasis more commonly seen in the lower extremities.

Tumor embolization (choice D) is incorrect. They are generally small but uncommon.

Vasculitis (choice E) is incorrect as this is not a surgical complication.
A 44 year-old female reports a waxing and waning cough, dyspnea and vague chest discomfort for the past five years. She also has intermittent fevers over the same time period with a six pound weight loss. On exam, red, tender subcutaneous swelling is found on both lower limbs, and decreased breath sounds and rales are found bilaterally. A biopsy of one of the skin lesions shows noncaseating granulomas. A chest X-ray is shown. Which of the following electrolyte abnormalities is most likely evident in this patient's plasma?

a. Hypercalcemia
b. Hyperkalemia
c. Hypernatremia
d. Hypochloremia
e. Hyponatremia

correct answer: A

The likely diagnosis is pulmonary sarcoidosis. The noncaseating granulomas are evident in the skin lesions of erythema nodosum and the chest X-ray shows hilar adenopathy. An increase in serum vitamin D levels is a result of hypersecretion by pulmonary macrophages, resulting in the body retaining calcium. Hypercalcemia and hypercalciuria are apparent on laboratory examination.
A 66-year-old man presents with new-onset weight gain, excessive sweating, and increased fatigue. His only significant past medical history is a bout of uncomplicated pneumonia several years ago. On physical examination, his voice sounds hoarse, which he attributes to his 2-packs per day smoking habit for the last 25 years, and there are purple markings over his abdomen. Dexamethasone suppression test is abnormal, bilateral adrenal CT scan is negative. Chest X-ray reveals a left lung mass adjacent to the hilum. Assuming that a lung neoplasm is responsible for his symptoms, what is this patient’s most likely prognosis?

a. Benign course  
b. Good prognosis with surgery  
c. Good prognosis due to low incidence of metastatic spread  
d. Poor prognosis due to likely metastatic spread to distant sites  
e. Prognosis comparable to squamous cell carcinoma of the lung

**Correct answer:** D

This patient’s 50-pack-year history of smoking puts him at risk for carcinoma of the lung. His signs and symptoms are consistent with paraneoplastic syndrome, commonly seen in the setting of small cell lung cancer. Small cell lung cancer carries the worst prognosis of all lung carcinomas due to its early metastatic spread to distant sites, including the brain, liver, bone, and adrenal glands. For this reason, surgery is not generally considered as effective treatment. Small cell lung cancer frequently involves hilar and mediastinal lymph nodes, which can be demonstrated on chest X-ray and manifest as hoarseness due to invasion or compression of the left recurrent laryngeal nerve by the expanding lymph nodes.

Choices A, B, and C are incorrect. Squamous cell carcinoma (Choice E) carries the best prognosis of all lung cancer for potential 5-year survival.
A 26 year female is brought into the emergency department by ambulance for acute exacerbation of her asthma. She has been hospitalized in the past for her asthma and once was intubated. On exam her temperature is 37.8 degrees, BP 110/60 HR 110/bpm and she is breathing at 28 breaths/minute. She is in moderate distress and unable to talk in full sentences. Diffuse expiratory wheezes are heard throughout the lungs. An ABG is taken. Which of the following values would most likely signal a need for intubation?

a. PCO2 21
b. PCO2 28
c. PH 7.39
d. PH 7.54
e. PO2 94

**Correct answer:** C

This patient is currently in respiratory alkalosis since she is in the middle of an asthma attack. In respiratory alkalosis, an ABG shows pH>7.40, pCO2<40 and often an elevated pO2, due to the rapid breathing rate that blows off the CO2 (which is main acid in the blood). The work of breathing during an asthma attack is increased to overcome the constricted airways leading to an increase of lactic acid from the various muscles involved in breathing. Intubation is usually avoided in asthmatics, but cannot be avoided if the breathing muscles become tired out. This "tiring out" is seen on ABG when the pH begins to normalize because this "normal" pH is a sign that the patient is unable to blow off the CO2 and the lactic acid is building up. Therefore, although a pH of 7.39 (choice C) is considered "normal," during an asthma attack it is a ominous sign that the patient is going into respiratory failure.

Options A, B, D, E are all values that could be seen during respiratory alkalosis consistent with an asthma attack. Although not ideal, they are signs that the patient is breathing appropriately for the situation and medical therapy can be continued without intubation.
A 65 year old male presents to his primary care doctor because he has been experiencing weakness over the past couple of weeks. The patient notes that the weakness is greatest in the mornings and he feels like he cannot move his arms or keep his head up. However, as the day goes on, he finds that his strength increases. On further questioning, he admits to a dry cough that is sometimes blood streaked and has lost 20 pounds over the past 4 months. He is a long time smoker who smoked 1-2 packs per day since the age of 18.

The doctor orders a chest x-ray which shows a 4-cm irregular nodule near the right mainstem bronchus.

What is most likely primary malignancy affecting this patient?

- a. Adenocarcinoma of the lung
- b. Adenocarcinoma of the colon
- c. Renal carcinoma
- d. Small cell carcinoma of the lung
- e. Squamous Cell Carcinoma of the lung

**Correct answer**: D

This patient is suffering from Lambert-Eaton myasthenic syndrome (LEMS) which is most often found in association with small cell carcinoma of the lung (option D). LEMS is similar to myasthenia gravis but it is caused by an antibodies against the pre-synaptic Ca++ channels. Unlike myasthenia gravis, LEMS improves over the course of the day or with repetitive activity. In addition small cell carcinoma is usually a centrally located tumor in the lung

Squamous cell carcinoma (option E) is a centrally located carcinoma associated with the paraneoplastic syndrome of an increase of PTrH (recombinant PTH). This leads to an increase of calcium, but weakness is not associated with this syndrome.

Adenocarcinoma (option A) of the lung is often peripherally located in the lung and is not associated with LEMS.

Both renal carcinoma (C) and colonic carcinoma (B) can metastasize to the lung. However, they are not associated with LEMS.
A 42-year-old woman with a history of rheumatoid arthritis and seasonal allergies comes to your clinic complaining of pain, erythema, and swelling "on the hard part of my ears." When questioned, she also describes a low-grade pain in her chest, over her costochondral joints. Which of the following is the most likely diagnosis?

- a. Ankylosing spondylitis
- b. Osteoarthritis
- c. Reiter syndrome
- d. Relapsing polychondritis
- e. Scleroderma

**Correct answer:** D

This patient likely has relapsing polychondritis, an autoimmune disorder that may be associated with other autoimmune diseases like rheumatoid arthritis or systemic lupus erythematosus. Patients may complain of bilateral swelling and pain of external ears, nasal cartilage, costochondral joints, and other arthralgias. Involvement of the larynx, trachea, and bronchi may also be reported. Occasionally, cartilage destruction may evolve into deformities like a saddle nose or sagging ears. Other organ systems that may become involved include the cardiovascular system, kidney, and skin.
A 38-year-old man is brought to the Emergency Room complaining of chronic fatigue. A complete workup reveals that this man has primary hypoaldosteronism secondary to Addison’s disease. Which of the following dermatologic changes would help to distinguish this disease from secondary hypoaldosteronism?

a. Desquamation of Palms and Soles
b. Greater than 6 Cafe au lait spots on body
c. Hyperpigmentation
d. Malar Rash on Face
e. Red macular rash of abdomen

**Correct answer:** C

Primary hypoaldosteronism due to Addison’s Disease results from adrenal cortical atrophy, and so lower total circulating levels of cortisol and aldosterone. Because of decreased negative feedback to the pituitary gland, circulating levels of ACTH are increased. You should know that ACTH is derived from the precursor molecule POMC, which is cleaved to form ACTH and MSH (Melanocyte Stimulating Hormone). MSH directly stimulates skin hyperpigmentation. Secondary hypoaldosteronism results from the decreased production of ACTH, accompanied by decreased MSH, and no hyperpigmentation.

Desquamation of the palms and soles (A) is most often associated with Syphilis, Group A beta Hemolytic Strep (Scarlett Fever) or Staphylococcus infections (Toxic Shock Syndrome, Scalded Skin Syndrome).

The presence of greater than 6 café au lait spots (B) (macules) on the skin suggests the Neurofibromatosis 1 (NF1) genotype.

A malar, butterfly shaped rash (D) is associated with Systemic Lupus Erythematosus specifically.

A red macular rash (E) on the abdomen has multiple causes, including Erythema Migrans or infection with Legionnaire’s Disease.
A 23-year-old man presents to his primary care physician complaining of an itchy rash. His girlfriend noticed a single red, round patch on his back one week ago. While he did not think much of it at the time, multiple red patches have cropped up around it since then. Review of systems is otherwise negative. He denies any recent use of medications. Physical exam reveals erythematosus macules and patches on his back, chest, and upper arms. There are no lesions on his face. What is the most likely etiology of this rash?

a. Autoimmune
b. Bacterial
c. Fungal
d. Hypersensitivity reaction
e. Viral

**Correct answer:** E

The rash of pityriasis rosea is a papulosquamous eruption that begins with the appearance of "herald patches." A generalized rash with multiple oval-shaped lesions proceeds to appear approximately 1 to 20 days after the herald patches in a classically described "Christmas tree" pattern. Pityriasis rosea commonly affects the trunk, upper arms, and thighs, and usually spares the face. Pruritus is the most common complaint, and the rash itself spontaneously resolves within 6 to 8 weeks without treatment. Treatment is purely symptomatic. Pityriasis rosea is thought to be related to the herpes virus type 7, and is not contagious.

Pityriasis rosea is attributed to a viral agent, not an autoimmune (Choice A), bacterial (Choice B), fungal (Choice C), or hypersensitivity reaction (Choice D) etiology. The malar rash of systemic lupus erythematosis is a classic example of a rash associated with an autoimmune etiology. It has a "butterfly" distribution over the face, spreading over the cheeks and bridge of nose. Rashes associated with bacterial infections, such as scarlet fever or toxic shock syndrome, present in the context of other symptoms, including fever and malaise. The herald patches of pityriasis rosea can be confused with that of ringworm, a fungal infection caused by Tinea corporis. These lesions are erythematous and annular in shape, but do not present as herald patches followed by a more generalized outbreak. Direct microscopy of skin scrapings with KOH preparation will reveal hyphae, and topical antifungals are used to treat ringworm. Hypersensitivity reactions can present with rash, and lesions may also be pruritic and painful. However, history will often include exposure to medications, such as sulfa drugs, or infection by herpes simplex virus.
A 34 year old female presents to her primary care physician with a variety of complaints that have started over the past couple of years and that seem to be increasing in severity. She states that a couple of winters ago, her fingers become painful and blue when she went outside without gloves. She has also had pain in her elbows and knees that sometimes responds to ibuprofen. Most recently she has notice a purple rash on her face most prominent above her eyes. She denies muscle weakness, difficulty swallowing, palpitations or fever.

On exam, she is in no distress with normal vitals signs. Her lung examination is notable for a rubbing sounds, but the rest of her exam is within normal limits.

Which of the following antibodies is most specific for her disease?

a. Anti-Jo Ab
b. Anti-SCL-70 Ab
c. Anti-U1-RNP Ab
d. Anti-centromere Ab
e. Anti-ds DNA Ab

correct answer: C

With the constellation of different symptoms, Raynaud’s phenomenon, arthralgias, heliotrope rash and pleuritic friction rub on exam, this patient is most likely is suffering from Mixed Connective Tissue Disease. This disease presents with various symptoms that are typical of other connective tissues diseases and is seen with a positive anti-U1-RNP antibody (option C).

Anti-Jo (option A) is typically seen in polymyositis which presents with proximal muscle weakness and heliotrope rash (in dermatomyositis), but not with Raynaud’s.

Anti-Scl-70 (option B) and Anti-centromere (option D) are seen in diffuse and CREST forms of scleroderma, respectively. However, the rash described is not typical.

Anti-ds DNA (option E) is associated with SLE which typically presents with a malar (butterfly) rash not the heliotrope rash described in the stem
A 62 year-old male suffers a heart attack, is resuscitated, and is finally stable. A few moments later, the patient's pO2 drops and his pulse is no longer discernible. The EKG indicates atrial fibrillation. A drug is administered and the action potential waveform is modified as illustrated above. Which drug was administered?

a. Flecainide  
b. Mexiletine  
c. Procainamide  
d. Sotalol  
e. Timolol

**Correct answer:** C

Only Na channel blockers (Class I Antiarrhythmics) will affect the duration of the action potential by modulating Phase 0 (as illustrated in the figure). Class Ia blocks fast-acting Na channels, slowing Phase 0 upswing and increases the AP duration. Class Ib drugs increase refractoriness (thereby shortening the AP potential). Class Ic drugs dramatically slow Phase 0, but do not increase the AP duration. Procainamide, the only Class Ia Antiarrhythmic in this list, is therefore the best choice.

Flecainide is a Class IC antiarrhythmic  
Mexiletine is a Class IB antiarrhythmic  
Sotalol is a Class III (K+ channel blocker)  
Timolol is a Class II (Beta-blocker)
It is determined that the best course of therapy for a 65-year-old male with repeated bouts of atrial fibrillation is rate control therapy. Administration of which of the following is most appropriate?

a. Adenosine  
b. Nesiritide  
c. Lidocaine  
d. Metoprolol  
e. Mexiletine

**Correct answer:** D  
Rate control therapy generally increases the PR interval. Beta blockers (metoprolol), cardiac glycosides (digoxin), and Class IV ARs (diltiazem, verapamil) all result in PR lengthening.

Lidocaine and mexiletine are Class Ib AR, which function by decreasing the action potential (AP) duration, but not by affecting the AV node (and thus the PR interval). Nesiritide is recombinant BNP used for decompensated congestive heart failure. Adenosine results in temporary AV block, but isn’t useful here.
A 56-year-old male is found to have elevated LDL and low HDL levels on his lipid panel. You begin treatment with niacin. A month later, the patient returns with complaints of frequent flushing and lightheadedness. This complication could have been avoided by also prescribing which of the following medications?

a. Amlodipine  
b. Aspirin  
c. Loratidine  
d. Octreotide  
e. Propranolol

**Correct answer: B**

Aspirin, which inhibits cyclooxygenase, can reduce prostaglandin levels thereby preventing flushing.

Niacin is an effective treatment for patients with hyperlipidemia. It acts by inhibiting lipolysis and thereby decreasing the levels of LDL. An added benefit of niacin is that it can also elevate HDL levels. One of the side effects of niacin is flushing which is thought to be mediated by prostaglandins. Aspirin, which inhibits cyclooxygenase, can reduce prostaglandin levels thereby preventing flushing. Other notable side effects of niacin include: hyperglycemia, hyperuricemia and increased incidence of peptic ulcers.

Amlodipine is a calcium channel blocker that also causes flushing as a side effect. Therefore, this would not be warranted in this patient.

Loratidine is a 2nd generation H1 antagonist that is often used to reduce the symptoms associated with allergies. This patient is not experiencing an allergic reaction.

Octreotide is a somatostatin analog that is used in the treatment of carcinoid syndrome. While flushing is one of the symptoms of carcinoid syndrome, this is a rare entity and not as likely given the other history in the question stem.

Propranolol is a non-selective beta blocker. One of the side effects of beta blockers is hyperlipidemia and this would therefore not be a desirable drug to use in this patient.
A patient with heart and renal failure presents to the emergency department complaining of a three-week history of palpitations and progressively deteriorating vision. Laboratory tests are notable for a potassium of 3.2 and a calcium of 11.0. Which of the following drugs most likely accounts for her symptoms?

- a. Captopril
- b. Digoxin
- c. Esmolol
- d. Nifedipine
- e. Verapamil

**Correct answer:** B

Digoxin is a cardiac glycoside classically used to treat congestive heart failure, but has been largely replaced in recent years due to its narrow therapeutic window. Xanthopsia (yellowing and/or blurring of vision) and cardiac dysrhythmias are common side effects of digoxin toxicity. The incidence of digoxin toxicity is elevated in patients with hypokalemia and hypercalcemia, as these conditions enhance the activity of digoxin.

Captopril is an angiotensin converting enzyme (ACE) inhibitor and is not associated with vision changes or cardiac dysrhythmias.

Esmolol is a beta-adrenergic receptor antagonist (beta blocker) used most frequently in emergency situations due to its rapid onset and short duration of action. While beta blockers can induce cardiac dysrhythmias, they are not associated with vision deterioration.

Nifedipine and verapamil are calcium channel blockers. They are commonly administered for hypertension and angina pectoris, not congestive heart failure.
At the onset of an acute myocardial infarction, a patient was placed on an intravenous vasodilator. Three days later, the patient had become short of breath, cyanotic, and occasionally drifted in and out of consciousness. Arterial blood gas results revealed an oxygen saturation of 87% on room air. The arterial blood was chocolate-brown in color. On what drug was the patient most likely placed?

a. Adenosine  
b. Digoxin  
c. Glyceryl trinitrate  
d. Nifedipine  
e. Verapamil

Correct answer: C

Glyceryl trinitrate, an organic nitrate, is often given IV at the onset of acute MI. A side effect of all organic nitrates is the oxidation of hemoglobin to form methemoglobin, which decreases the oxygen carrying capacity of blood. This produces shortness of breath, cyanosis, and can occasionally cause unconsciousness. Blood color is characteristically chocolate-brown or dark red.
A 42 year old male with a past medical history of diabetes mellitus is found during a regular physical to have a BP of 145/85. A repeat BP taken 2 days later shows 143/88. What is the most appropriate initial therapy?

a. Enalapril
b. Furosemide
c. Hydrochlorothiazide
d. Metoprolol
e. Nifedipine

**Correct answer:** A

This patient is suffering from hypertension and has a history of diabetes which are indications for treatment. In patients with diabetes and hypertension the first line agent is an ACE inhibitor like enalapril (option A) because it has renoprotective effects in addition to its blood pressure reducing qualities.

Furosemide is a diuretic that can be used to treat hypertension but it is not a first line agent.

Hydrochlorothiazide is a first line agent for the treatment of hypertension in non-diabetic patients.

Metoprolol is a B-blocker that is used for the treatment of hypertension but is not a first line treatment. Also, B-blockers are contraindicated in diabetic patients due to its potential to mask signs of low blood sugar.

Nifedipine is a Calcium channel blocker that is used in the treatment of hypertension but it is not a first line therapy.
A 58-year-old man is being treated for hypertension. His internist prescribes lisinopril. Medications of this class should be avoided in most patients with which of the following conditions?

a. Congestive heart failure  
b. Myocardial infarction  
c. Chronic renal insufficiency  
d. Bilateral renal artery stenosis  
e. Diabetes mellitus

**Correct answer:** D

Lisinopril is an angiotensin converting enzyme inhibitor (ACEI). ACEIs reduce blood pressure by inhibiting the production of angiotensin II (AngII), which decreases vascular smooth muscle tone. Patients with bilateral renal artery stenosis, however, rely on AngII to increase resistance in the postglomerular arteriole in order to maintain adequate glomerular filtration. Blocking this response may tip them into acute renal failure.

The first three answer choices are specific indications for the use of ACEIs. ACEIs provide mortality benefits following myocardial infarction and in patients with congestive heart failure. They have also been shown to offer renal protection in patients with chronic renal insufficiency. There is mounting evidence that ACEIs are beneficial in diabetics as well.
A 50-year-old male begins treatment with an antiarrhythmic medication for recurrent atrial fibrillation. The arrhythmia resolves and there is now mild bradycardia and significant QT interval prolongation on his ECG recording. Which of the following drugs was most likely used in this patient?

a. Metoprolol  
b. Propranolol  
c. Sotalol  
d. Carvedilol  
e. Verapamil  
f. Diltiazem

**Correct answer:** C

1. The QT-interval represents the period of ventricular repolarization as the ventricles prepare for the next beat. Repolarization is mediated by K⁺ efflux from the cardiac myocyte. Sotalol is a beta-blocker with class 3 (K⁺ channel blocking) antiarrhythmic properties. Other class 3 antiarrhythmics include amiodarone, ibutilide, and dofetilide. Sotalol is the only class 3 antiarrhythmic with beta-adrenergic blocking abilities (causing the patients mild bradycardia) as well as class 3 effects (causing the QT interval prolongation).

(Choices A, B, and C) Metoprolol, propranolol, and carvedilol are beta-blockers like sotalol, but these medications lack sotalol's class 3 K⁺ blocking effects. Beta-blockers decrease heart rate and cardiac contractility by slowing AV nodal conduction and the phase 4 depolarization of cardiac pacemaker cells. They are useful as rate controlling agents, as myocardium protective agents following myocardial infarction or in CHF resulting from systolic dysfunction, and as antihypertensives. They do not prolong the QT interval.

(Choices E and F) Verapamil and diltiazem are calcium channel blockers that affect cardiac tissue and are therefore class 4 antiarrhythmic agents. Verapamil is the most cardioselective, but both will act on the peripheral vasculature as well, causing a decrease in blood pressure. As calcium channel blockers, these medications slow the phase 0 depolarization of cardiac pacemaker cells and phase 2 of the myocyte action potential, neither of which will change the QT interval. Their adverse effects include negative inotropy and the potential for AV block. Verapamil is additionally associated with gingival hyperplasia and constipation.

Educational Objective: Sotalol has both beta-adrenergic blocking properties and class 3 antiarrhythmic (K⁺ channel blocking) properties. It prolongs both the PR interval and the QT interval.
Baby Aspirin is a drug used as an antiplatelet, working by inhibiting Cyclooxygenase pathway, knowing that it blocks both Thromboxane A2 (increases platelets aggregation) and Prostacyclin PGI2 (decreases platelets aggregation). The reason behind baby Aspirin being an antiplatelet without affecting the needed function of Prostacyclin is:

- a. Prostacyclin can be replaced by other pathway
- b. Small amount of aspirin is enough to block Thromboxane synthesis, because Platelet doesn't have a nucleus
- c. Thromboxane A2 blockade is irreversible, while Prostacyclin blockade is reversible
- d. The inhibition of prostacyclin is not important in the pathophysiology of thrombos formation
- e. Unknown reason

**Correct answer:** B

Aspirin taken in small doses inhibits the Cyclooxygenase pathway, so you think it inhibits both Prostacyclin and Thromboxane A2 equally, but it has a greater effect on Thromboxane over Prostacyclin, because platelet doesn't have a nucleus, so the low dose of aspirin is enough to inhibit Thromboxane synthesis, while endothelial cell, the source of Prostacyclin has a nucleus, and can overcome the low dose of aspirin, and continue synthesising Prostacyclin.
A 58 year old man with a past medical history of hypertension presents with a cough. The medical team suspects that the cough is due to one of his antihypertensive medications. Which of the following is a characteristic of the best alternative drug for this patient?

- a. Interference with binding of angiotensin I
- b. Interference with binding of angiotensin II
- c. Reduction of serum angiotensin II levels
- d. Decreased production of a key enzyme produced in the lungs
- e. None of the above

correct answer: B

This patient’s cough is side effect of treatment by ACE inhibitor (drug with an -opril suffix, such as captopril). The best alternative is an angiotensin receptor blocker (ARB), which treats hypertension without causing cough. ARBs (such as losartan) work by inhibiting binding of angiotensin II to its receptor, thereby reducing the production of aldosterone.

A is incorrect because angiotensin I binds to angiotensin converting enzyme (ACE) leading to formation of angiotensin II

C is incorrect because angiotensin II levels are unaffected. In fact, the amount of free angiotensin II may actually increase from lack of access to its binding sites, which are occupied by the drug.

D is incorrect because ACE is produced in the lungs. Its production is reduced by ACE inhibitors, which are the cause of this patient’s presentation.
Which of the following is correct about Warfarin?

a. warfarin is a rat poison
b. Maximum dosage is 13 mg
c. warfarin is not an oral anticoagulant
d. It is useful in pregnancy to prevent thrombosis
e. It is used to prevent heart attack

**correct answer:** B

Warfarin is not a rat poison. Maximum dosage is 13 mg to prevent stroke after a thromboembolic disorder. It is an oral anticoagulant. It is not useful in pregnancy as it causes craniofacial abnormality in newborn. Its maximum dosage is 13 mg and it differs in each person. Over dosage causes bleeding. Minimum amount is 3 mg. It's dosage differs on each person according to their genetic variation.
A 50 year old woman with a history of headache presents with complaints of chest pain. She described the pain as pressure, and tightness in her chest. She said that the pain occurred after taking a drug, the name of which she forgot. The most likely mechanism of this drug is:

- a. Antagonist action at serotonin receptor
- b. Agonist action at serotonin receptor
- c. Release of nitric oxide
- d. Inhibition of calcium channel opening
- e. Increased synaptic levels of catecholamines

**Correct answer:** B

This patient is most likely to have taken Sumatriptan, a drug that is used in the treatment of migraine headaches. Sumatriptan is a selective serotonin (5 HT1) receptor agonist (A is incorrect) that may have the dangerous side effect of coronary vasospasm, which can cause chest pain due to myocardial ischemia.

C is incorrect. A drug that causes the release of nitric oxide would cause vasodilatation, and improve perfusion of the heart. That clearly would not cause ischemia.

D is incorrect. Calcium channel blockers are also used in the treatment of migraine headache and heart disease. They also lead to vasodilatation, enhancing perfusion of the heart, and therefore would not cause chest pain due to ischemia.

E is incorrect. Cocaine is another classic drug that may cause coronary vasospasm. It stimulates presynaptic release of catecholamines (norepinephrine and dopamine) and inhibits catecholamine reuptake, causing increased levels at the synapse.
Mean arterial blood pressure (MAP) and heart rate (HR) measurements were recorded during the intravenous administration of two different drugs. Select the most likely drugs given at the indicated points:

a. X, Acetylcholine Y, phentolamine
b. X, epinephrine Y, hexamethonium
c. X, isoproterenol Y, propranolol
d. X, metaproterenol Y, propranolol
e. X, norepinephrine Y, hexamethonium

**correct answer:** E

The correct answer is E. There are several strategies that are useful in solving drug tracing problems. First, always look at blood pressure first and heart rate second. Blood pressure changes will be the direct result of the administered drug; heart rate changes may be a direct effect or they may result from a baroreceptor reflex. Second, assume that when an agonist effect is gone, so is the agonist; however, assume that an antagonist stays on board for the entire trace. Third, begin by eliminating as many options as possible by examining the initial agonist effect.

The only two agonists that would be expected to raise blood pressure to this degree are norepinephrine (alpha-1, alpha-2, and beta-1 agonist) and phenylephrine (alpha-1 agonist). Thus, choices A, B, C, and D can be eliminated immediately.

Drug Y lowers blood pressure and raises heart rate. Hexamethonium (ganglionic blocker) would lower blood pressure and increase heart rate by blocking the predominant tone of the arterioles (sympathetic) and the heart (parasympathetic). Phentolamine (alpha antagonist) would lower blood pressure by blocking arteriolar alpha-1 receptors. The increase in heart rate would be a baroreceptor reflex.

The second administration of Drug X causes an increase in both blood pressure and heart rate. Only the combination of norepinephrine (NE) and hexamethonium could do this. NE would still increase blood pressure by stimulating end organ receptors; however, the baroreceptor reflex would be blocked by hexamethonium. NE’s ability to increase heart rate by stimulating cardiac beta-1 receptors is now revealed.

Choice A) Acetylcholine would cause a decrease in blood pressure by stimulating non-innervated muscarinic receptors present on arterioles. Heart rate would increase because of a baroreceptor reflex. Phentolamine would cause a decrease in blood pressure and an increase in heart rate (baroreceptor reflex). It should have no effect on a subsequent ACh administration.

Choice B) Epinephrine would cause a small increase in mean blood pressure, but not enough to elicit a baroreceptor reflex. So, an increase in heart rate would be seen as a result of beta-1 stimulation of the heart. Hexamethonium would cause a decrease in blood pressure and an increase in heart rate. Hexamethonium should not affect a subsequent epinephrine administration because a baroreceptor reflex was not initially produced.

Choice C) Isoproterenol (beta-1 and beta-2 agonist) would cause a decrease in blood pressure (by stimulating arteriolar beta-2 receptors) and an increase in heart rate (a combination of cardiac beta-1 receptor stimulation and baroreceptor reflex). Propranolol (beta-1 and beta-2 antagonist) would decrease blood pressure and heart rate. It would completely block a subsequent administration of isoproterenol.

Choice D) Metaproterenol (beta-2 agonist) would decrease blood pressure (by stimulating arteriolar beta-2 receptors) and increase heart rate (baroreceptor reflex). Propranolol (beta-1 and beta-2 antagonist) would decrease blood pressure and heart rate. It would completely block a subsequent administration of metaproterenol.

Choice F) Phenylephrine would increase blood pressure and decrease heart rate (baroreceptor reflex). Phentolamine would decrease blood pressure and increase heart rate (baroreceptor reflex). Phentolamine would completely block a subsequent administration of phenylephrine.
You decide to prescribe a statin to a 42-year-old female patient who has had repeatedly elevated cholesterol levels despite total lifestyle changes. She asks you how this class of drug works. What could you tell her?

- a. Statins inhibit the rate-limiting step of cholesterol synthesis.
- b. Statins inhibit HMG-CoA reductase.
- c. Statin inhibit the synthesis of mevalonic acid.
- d. Choice A or B.
- e. All of the above.

**Correct answer:** E

HMG-CoA reductase converts 3-hydroxy-3-methylglutaryl-CoA (HMG-CoA) to mevalonic acid as part of the mevalonate pathway. It is the rate-limiting step of de novo cholesterol synthesis.
A 43-year-old woman presents with bone pain, joint pain, and fatigue. Further history reveals blood in her urine and difficulty moving her bowels. She denies recent weight loss, poor appetite, and night sweats. She has no chronic medical conditions and does not take any medications or use drugs. Her family history is negative for malignancy. Physical examination is negative for lymphadenopathy or hepatosplenomegaly. Laboratory values include serum calcium 11.2 mg/dL, serum phosphorous 2.0 mg/dL, urine calcium 410 mg, and increased levels serum 1,25-hydroxy Vitamin D, and parathyroid hormone. Which of the following medications would most likely help in treating this patient?

a. 1,25-hydroxy Vitamin D
b. Calcium gluconate
c. Furosemide
d. Hydrochlorothiazide
e. Magnesium sulfate

correct answer: C

This patient presents with signs of hypercalcemia, or “stones, bones, groans, moans, and overtones.” These include kidney stones, nephrocalcinosis, thirst, polyuria, bone pain and fractures, anorexia, dyspepsia, constipation, fatigue, myalgia, proximal muscle weakness, joint pain, depression, memory loss, confusion, lethargy, and coma. Several etiologies may result in hypercalcemia, including primary hyperparathyroidism, multiple endocrine neoplasia syndromes, exogenous vitamin D administration, and malignancy. In primary hyperparathyroid, serum calcium, 1,25(OH)2D, and parathyroid hormone are elevated while serum phosphorous is low. Urine calcium is also elevated.

Furosemide (Choice C) is a loop diuretic that inhibits the cotransport of Na+/K+/2Cl- in the ascending limb of the loop of Henle, thus decreasing reabsorption and increasing excretion of these ions and Ca2+

1,25(OH)2 Vitamin D (Choice A), or Vitamin D, is used in the treatment of hypocalcemia. It increases intestinal calcium and phosphate absorption while decreasing renal excretion, thus increasing the blood levels of both ions.

Calcium gluconate (Choice B) is used in the treatment of hypocalcemia. It would be contraindicated in this case as the patient’s serum calcium is already high.

Hydrochlorothiazide (Choice D) is a thiazide diuretic that can be used in the treatment of hypocalcemia. In contrast to furosemide, it promotes the reabsorption of calcium to decrease the calcium content of urine.

Magnesium sulfate (Choice E) is used in the treatment of hypomagnesemia, which is often associated with hypocalcemia.
A 35-year-old female complains of a white colored discharge from her nipples. Upon further questioning, she admits that she has not had her period for the past six months and she has found herself tripping more often in her apartment. On physical examination, it is noted that her peripheral vision is impaired. What medical therapy would most likely help with her symptoms?

- a. Bromocriptine
- b. Leuprolide
- c. Levothyroxine
- d. Prednisone
- e. Propylthiouracil (PTU)

**Correct answer: A**

This patient is suffering from a prolactinoma. Hyper secretion of prolactin can stimulate galactorrhea which causes the milky discharge from nipples, suppression of ovulation, and infertility (not mentioned in this question but a common sign). In addition, since this tumor is located in the pituitary, if the tumor is large enough, it can press on the the optic chiasm and causes bitemporal hemianopsia causing loss of peripheral vision. Prolactin secretion is suppressed by dopamine and bromocriptine (option A) is a dopamine analog.

Leuprolide (option B) is a GnRH analog that can be used to suppress the hypothalamic-pituitary axis in conditions such as prostate cancer (medical castration)

Levothyroxine (option C) is a synthetic analog of T4 (thyroid hormone) which is used hypothyroidism. Although galactorrhea and anovulation can be seen in severe hypothyroidism (since TRH can also stimulate prolactin), hypothyroidism would not cause loss of peripheral vision.

Prednisone (option D) is a glucocorticoid and used for many purposes but not useful in the case of a prolactinoma.

PTU (option E) is used in hyperthyroidism to suppress thyroid function.
A 25-year-old male patient complains of increased thirst and increased frequency of urination. He denies any trauma or recent surgery. At baseline his urine specific gravity is 1.001 and his serum osmolality is 350.

He is admitted to the hospital for observation and water restriction. After 6 hours, his urine specific gravity is 1.003 and his serum osmolality is 365.

He is given intranasal desmopressin (DDAVP) and water is restricted for 6 hours. Afterward his urine specific gravity is 1.004 and his serum osmolality is 400.

What medication most likely caused these symptoms?

- a. Aspirin
- b. Furosemide
- c. Gentamicin
- d. Lithium
- e. Penicillin

Correct answer: D

This patient is suffering from nephrogenic diabetes insipidus which is a side effect of lithium (option D). Diabetes insipidus is characterized by increased thirst (polydipsia) and increased urination (polyuria) with the inability to concentrate urine due to a lack of ADH. Therefore, urine specific gravity will be low (<1.006) and serum osmolality will be >250. Nephrogenic diabetes insipidus (lack of response to ADH) can be distinguished from central (loss of ADH production) because nephrogenic DI will not respond to DDAVP which is a ADH analog.

Furosemide (option B) is a loop diuretic which would cause decreased urine specific gravity but the serum osmolality would not increase as much.

Aspirin (option A), Gentamicin (option C) and Penicillin (option E) can cause acute tubular necrosis (ATN) and acute interstitial nephritis which would cause an increase in serum creatinine but would not cause an low urine specific gravity or a high serum osmolality.
What is the mechanism of action of Allopurinol?

a. Inhibits uric acid formation.
b. Increases xanthine oxidase synthesis.
c. Binds to tubulin.
d. Increases purine synthesis.
e. Inhibits cyclooxygenase.

**Correct answer:** A

This drug inhibits uric acid formation (answer A). Allopurinol is an isomer of hypoxanthine (a naturally occurring purine in the body) and it inhibits xanthine oxidase (answer B). This enzyme is responsible for the successive oxidation of hypoxanthine and xanthine resulting in the production of uric acid, the product of human purine metabolism. Allopurinol decreases both uric acid formation and purine synthesis. (answer D).

Colchicine binds to tubulin (answer C) and Indomethacin inhibits cyclooxygenase (answer E).
Basic science: Pharmacology

Question: 766

Clinical science: Endocrine

Bevacizumab can be used to treat which complication of Diabetes mellitus

a. Neuropathy
b. retinopathy
c. nephropathy
d. ketoacidosis
e. non alcoholic steatohepatitis

Correct answer: B

This drug blocks VEGF, an important mediator of proliferative retinopathy.
A 46 year old man is about to undergo surgery to remove a pheochromocytoma. Which of the following drugs (with its correct mechanism of action) is most likely to have been administered prior to surgical treatment?

a. phenylephrine: alpha receptor agonist (nonspecific)
b. phenylephrine: alpha receptor antagonist (nonspecific)
c. prazosin: alpha receptor antagonist (nonspecific)
d. prazosin: alpha-1 selective receptor antagonist
e. phenoxybenzamine: alpha receptor antagonist (nonspecific)

correct answer: E

Pheochromocytomas are tumors that secrete large amounts of catecholamines, including norepinephrine, epinephrine, and dopamine. They can cause a range of symptoms including hypertension, palpitations, diaphoresis, and anxiety. Before surgery, the patient should be given phenoxybenzamine, an irreversible nonspecific alpha receptor antagonist, in order to fully block the alpha receptor effects of these catecholamines.
All the following medications have been found to induce high levels of glucose, except:

a. Prednisone
b. Pentamidine
c. Thiazides
d. ACE inhibitors
e. alpha-interferon

**Correct answer:** D

ACE inhibitors are not known to cause diabetes mellitus.

Glucocorticoids, pentamidine, thiazide diuretics, and alpha interferon can cause Diabetes mellitus.
A 52-year-old man with recently diagnosed type 2 diabetes mellitus comes to the physician for a follow-up examination. Physical examination shows no abnormalities. Laboratory studies show an increased hemoglobin A1c despite patient compliance with diet and exercise recommendations. Treatment with a sulfonylurea is started. Which of the following is most likely to occur in this patient?

- a. Decreased production of glucose from the liver
- b. Decreased secretion of insulin from the pancreas
- c. Decreased speed of carbohydrate absorption from the intestines
- d. Increased production of glucose from the liver
- e. Increased secretion of insulin from the pancreas

**Correct answer:** G

Sulfonylureas increase the secretion of insulin from pancreatic beta cells (choice E) by binding to ATP-dependent potassium channels causing an inhibition of the outflux of potassium. This causes a depolarization of the cell membrane, which opens voltage-dependent calcium channels and stimulates secretion of insulin-containing granules. In contrast, sulfonylureas do not decrease the secretion of insulin from the pancreas (choice B).

Changing the amount of glucose produced by the liver (choices A and D) is also not the mechanism of action of sulfonylureas. Metformin is an example of a drug that can decrease the amount of glucose produced by the liver.

Sulfonylureas also do not decrease the speed of carbohydrate absorption from the intestines (choice C).
A 46 year old diabetic woman presents with intense nausea for 2 weeks duration. She is subsequently diagnosed with diabetic gastroparesis and started on metoclopramide as an anti-emetic. Which of the following drugs has a similar mechanism of action as that of metoclopramide?

a. Prochlorperazine  
b. Dronabinol  
c. Aprepitant  
d. Ondansetron  
e. Cisapride

**Correct answer:** A

Metoclopramide is a D2 receptor antagonist that increases gastrointestinal contractility and motility as well as increases tone in the lower esophageal sphincter. Prochlorperazine (choice A) is also a D2 receptor antagonist and is used as an anti-emetic.

Dronabinol (choice B) is a cannabinoid and is used as a centrally acting anti-emetic.

Aprepitant (choice C) is a centrally acting anti-emetic that selectively antagonizes human substance P and neurokinin 1 receptors. Ondansetron (choice D) is a centrally acting anti-emetic that selectively antagonizes 5-HT3 receptors.

Cisapride (choice E) acts through serotonin receptors to increase ACh release in the myenteric plexus, promoting gastric motility. It is currently banned from U.S. markets due to its toxicity of cardiac arrhythmias.
A 58 year old alcoholic man presents with diarrhea for 4 days duration. He further explains that for the past week he has been taking “heartburn medication” for worsening epigastric pain. Blood pressure was 100/60 with pulse rate at 95 bpm. Laboratory studies were all normal except serum potassium of 3.3 mEq/L. Which of the following medications did the patient most likely overuse?

a. Cimetidine
b. Aluminum hydroxide
c. Calcium carbonate
d. Magnesium Hydroxide
e. Ondansetron

**Correct answer**: D

Alcoholics commonly have gastritis for which they seek relief with over-the-counter medications such as antacids. There are various antacids in the answer choices, each with their own side-effect profile. Magnesium hydroxide (choice D) is the only medication listed that has an overuse effect of diarrhea and hypotension. Additional side-effects include hyporeflexia, hypokalemia, and cardiac arrest.

Cimetidine (choice A) is a H2 receptor blocker with side effects of gynecomastia, impotence, dizziness, and headaches.

Aluminum hydroxide (choice B) is an antacid with side effects of constipation, hypophosphatemia, proximal muscle weakness, osteodystrophy, seizures, and hypokalemia.

Calcium carbonate (choice C) is an antacid with side effects of hypercalemia, rebound acid secretion, and hypokalemia.

Ondansetron (choice E) is a centrally acting anti-emetic selectively antagonizing 5-HT3 receptors. Side effects include headache and constipation.
A 55-year-old man presents to the emergency room with greasy, foul-smelling diarrhea for the past 4 weeks. He is a long time alcoholic and has been hospitalized several times in the past for a "stomach problem." He denies having had fever, hematochezia, or mucus in his stools. Which medication is most likely to benefit the patient at this time?

a. Esomeprazole  
b. Metronidazole  
c. Pancreatic enzymes  
d. Sulfasalazine  
e. Vancomycin

**correct answer: C**

This patient is likely suffering from pancreatic insufficiency secondary to chronic pancreatitis caused by alcoholism. Pancreatic enzymes will help him digest and absorb fats.

There is no evidence of bacterial infection that would warrant antibiotics (B,E).
There is no evidence of colonic inflammation or systemic symptoms that would indicate inflammatory bowel disease (D).
This patient's symptoms are not typical of GERD, gastritis, or PUD (A).
A 14 year old male patient presents with altered mental status, and RUQ pain. On physical exam, brown corneal deposits are noted on slit-lamp exam. Hepatomegaly is noted with a span 12cm, and his asterixis is noted. Laboratory values are notable for elevated transaminases.

What is most appropriate medical therapy?

a. D-penicillamine  
b. Deferoxamine  
c. Dimercaprol  
d. N-acetylcysteine  
e. Therapeutic phlebotomy

**Correct answer: A**

This patient is suffering from Wilson's Disease. The signs of Wilson's are Kayser-Fleischer rings (brown rings around iris), and liver disease which can progress to cirrhosis. Wilson's disease is caused by the accumulation of copper which is treated with d-penicillamine (option A).

Deferoxamine is used for the treatment of increased iron usually used in those with multiple blood transfusions. Therapeutic phlebotomy (option E) is used in hemochromatosis which is also caused by the build up of iron.

Dimercaprol (option C) is used to treat heavy metal poisoning, and is only the last line of defence in Wilson's disease.

N-acetylcysteine is used for acetaminophen overdoses.
A 67-year-old woman is started on warfarin to prevent embolic events from her atrial fibrillation. Four days after taking warfarin, she starts developing well defined, erythematous, indurated, and purpuric lesions on her thighs. The inhibition of which component of the coagulation cascade is likely responsible for these lesions?

a. Factor II  
b. Factor VII  
c. Factor IX  
d. Protein C  
e. Protein S

**Correct answer:** D

Warfarin is an oral anticoagulant that interferes with normal synthesis of vitamin K-dependent clotting factors in the liver. Clotting factors II, VII, IX, and X are inhibited by warfarin, leading to an increase in prothrombin (PT) time and international normalized ratio (INR). Proteins C and S are also inhibited by warfarin; however they normally function as anti-coagulants.

Protein C (choice D) is an anti-coagulation factor with a very short half-life (7.9 hours), so its level can decrease first. This may lead to the paradoxical pro-coagulant effect of warfarin seen here, called warfarin necrosis.

Protein S (choice E) is also an anti-coagulation factor, but it has a longer half-life than protein C.

Factors II, VII, and IX (choices A, B, C) are pro-coagulation factors that warfarin inhibits; thus, hemorrhage and easy bruising would be resultant side effects. Factor VII has the shortest half life (3-6 hours).
After taking a new prescription medication for a urinary tract infection, a 17-year-old Greek male presents with an elevated bilirubin and a hemoglobin of 10. He is diagnosed with an enzyme deficiency of the pentose phosphate pathway. The medication most likely to have caused his symptoms has which of the following mechanisms of action?

a. Binds to the bacterial 30S ribosomal subunit
b. Competitive antagonist of para-aminobenzoic acid
c. Cyclooxygenase inhibitor
d. Inhibitor of DNA gyrase
e. Inhibitor of vitamin K epoxide reductase

correct answer: B

This patient suffers from G6PD deficiency, an enzyme disorder common among people of Mediterranean descent that leads to hemolytic anemia during episodes of oxidative stress. The patient most likely took trimethoprim-sulfamethoxazole for his UTI; Trimethoprim acts by interfering with the action of bacterial dihydrofolate reductase, inhibiting synthesis of tetrahydrofolate acid. Tetrahydrofolate acid is an essential precursor in the de novo synthesis of the intermediate Thymidine monophosphate (dTMP), precursor of DNA metabolite Thymidine triphosphate[1]. Bacteria are unable to take up folic acid from the environment (i.e. the infection host) and are thus dependent on their own de novo synthesis. Inhibition of the enzyme starves the bacteria of nucleotides necessary for DNA replication. Sulfamethoxazole is a competitive antagonist of para-aminobenzoic acid in the folate synthesis pathway (choice B) and leads to oxidative stress and hemolytic anemia in patients with G6PD deficiency.

The other drugs mentioned are not known to cause hemolytic anemia in G6PD deficiency.

Aminoglycosides bind to the bacterial 30S ribosomal subunit (Choice A) - not first line for treating UTI.

The cyclooxygenase inhibitor (choice C) aspirin is also not likely to be prescribed for a UTI.

Quinolones act as inhibitors of DNA gyrase (choice D) but are also not first line for UTIs.

An inhibitor of vitamin K epoxide reductase (Choice E) is coumadin, an inappropriate choice for treating a UTI.
A patient is administered a pharmacologic agent topically onto the eye, resulting in mydriasis without cycloplegia. Which of the following agents was administered?

a. Atropine  
b. Bethanechol  
c. Neostigmine  
d. Phenylephrine  
e. Pralidoxime

**Correct answer:** D

The anterior chamber of the eye is controlled primarily by three muscles (pupillary constrictors, pupillary dilators, and ciliary muscle). Activation of constrictors (through muscarinic receptors) leads to miosis/constriction; activation of dilators (through alpha-adrenergic receptors) leads to mydriasis/dilation; and activation of the ciliary muscle (through muscarinic receptors) allows accommodation. Bethanechol and neostigmine result in increased ACh & muscarinic activation, thus causing miosis in the pt. Atropine would block all muscarinic activation, resulting in not only mydriasis but also cycloplegia (loss of accommodation due to ciliary muscle paralysis). Pralidoxime is an ACh reactivator, so it wouldn't have any effect here. Therefore, phenylephrine—a selective alpha1-adrenergic agonist—is the only viable possibility.
A patient self-administers a high, but non-toxic dose of a first-generation antihistamine. Which of the following symptoms is likely to be found in this patient?

a. Blurred vision  
b. Hyperactivity  
c. Increased blood pressure  
d. Nausea  
e. Parkinsonism

**Correct answer:** A

Anticholinomimetic actions are common side-effects of non-selective H1 antagonists. This can result in blurred vision and urinary retention.

H1-antagonists at high, non-toxic doses cause:

1. central sedation (i.e. drowsiness with diphenhydramine), not hyperactivity;
2. adrenergic-blocking (in high doses), so hypotension, not hypertension;
3. anti-emetic action, so no nausea; and
4. anti-cholinergic effects, so antiparkinsonism
A 25 year old from Massachusetts complains of increasing fatigue and pre-syncope. Upon further questioning, he admits to having had a rash on his arm a couple of months ago. He denies joint pain, photophobia or a stiff neck.

An EKG is taken which shows 3rd degree heart block (complete heart block). His laboratory tests are within normal limits. Blood cultures are pending.

What is the most appropriate treatment?
   a. Ceftriaxone
   b. Doxycycline
   c. Implantable defibrillator
   d. Penicillin G
   e. Rifampin

**Correct answer:** A

This patient is suffering from Lyme disease which is endemic to the North-East of the United States. It is borne by a tick and caused by the spirochete Borrelia burgdorferi. After the tick infects the patient, many patients experience a rash that has a target appearance (erythema migrans). Lyme disease in later stages can cause meningitis, arthritis and heart block. The most appropriate treatment is ceftriaxone, which is the drug of choice when the patient develops advanced signs of lyme disease, that include Complete heart block or CNS involvement.

Doxycycline (Choice B), a tetracycline which is the primary treatment for stage 1 of Lyme disease.

Although this patient is found to be in heart block, the most appropriate treat at this moment is doxycycline rather than an implantable defibrillator (Choice C) since the heart block will improve with treatment.

Penicillin G (Choice D) given in high doses can be use in the treatment for cardiac complications of Lyme Disease.

Rifampin (Choice E) is part of TB treatment.
A boy is born at 40 weeks gestation with APGAR scores of 9 and 9 at 1 and 5 minutes. It is noted in the well baby nursery that this infant is breathing faster than the other newborns. On physical exam, his heart rate is 153 and he has a murmur that is heard throughout both systole and diastole. Over the next couple of days, the newborn continues to breathe rapidly and the murmur does not change. An echocardiogram shows no congenital heart anomaly. What medication is contraindicated for this newborn?

- a. Acetaminophen
- b. Ampicillin
- c. Indomethacin
- d. Prostaglandin E1
- e. Vitamin K

**Correct answer:** D

This infant has a patent ductus arteriosus (PDA). During gestation, the ductus arteriosus is a shunt from the pulmonary artery to the aorta that helps bring oxygenated blood to the body. Upon delivery, the infant starts to breathe and systemic resistance becomes greater than pulmonary resistance and the shunts begin to close (foramen ovale, ductus venosus, ductus arteriosus). If the ductus arteriosus remains open after birth blood is shunted from systemic to pulmonary circulation and less oxygenated blood goes to the body leading to increased respirations (tachypnea). A PDA results in a machine-like murmur, continuous throughout systole and diastole. In this case, there is no congenital anomaly of the heart (transposition of the great vessels, tricuspid atresia, severe tetralogy of Fallot) and therefore, it would be best to assist with the closure of the PDA.

Prostaglandin E2 (option D) will help keep the PDA open, which is undesirable in this case. Note that the question asks which drug is contraindicated, so this is the correct answer.

Indomethacin (option C) is an NSAID and inhibits prostaglandin synthesis and therefore can be used to help close the PDA.

Ampicillin (option B) is an antibiotic that is often used to treat group B streptococcus infection in neonates. It does not affect the PDA and is safe.

Acetaminophen (option A) is safe to use in newborns for fever and pain.

Vitamin K (option E) is routinely given to newborns to prevent against hemorrhage since the newborn is vitamin K deficient and therefore can not produce adequate coagulation factors.
A 35-year-old woman is found unconscious in her apartment next to an empty bottle of pills. She is brought to the Emergency Department and given flumazenil, and shortly thereafter regains consciousness. Which of the following drugs did she likely ingest?

- a. Diazepam
- b. Eszopiclone
- c. Hydromorphone
- d. Oxycodone
- e. Zolpidem

**Correct answer: A**

Flumazenil is a benzodiazepine analog that competitively inhibits the actions of benzodiazepines, like diazepam (choice A). It can reverse benzodiazepine overdose, but there is a risk of withdrawal seizures.

Eszopiclone (choice B) and zolpidem (choice E) are nonbenzodiazepine sedative-hypnotics.

Hydromorphone (choice C) and oxycodone (choice D) are opioids that bind to the mu receptor.
A 42-year-old woman, originally from Jamaica, with rheumatoid arthritis presents with a persistent cough. Her chest X-ray is above. Which of the following drugs was she most likely taking for her rheumatoid arthritis?

a. Aspirin
b. Azathioprin
c. Etanercept
d. Indomethicin
e. Methylprednisolone

**Correct answer:** C

The chest x-ray is classic for active secondary tuberculosis (cavitary upper-lobe lesion).

Etanercept is a TNF-alpha antagonist. The other drug in this class is infliximab. Reactivation of latent TB infection is possible with this class of drugs because TNF-alpha inhibits macrophages, which are necessary for the control of latent mycobacterial infection.

The other drugs could also lead to immunosuppression and thus TB reactivation, but this adverse reaction is more commonly associated with the TNF antagonists because of the specificity of the immune suppression.
A 70 year old woman who underwent an aortic valve replacement two years ago is taking warfarin. For the past 2 years she has had no complications. Last week, after experiencing some depression, she noticed that the INR on her home INR monitoring kit read a value of 0.5. Which of the following is she likely taking in addition to warfarin?

a. Cimetidine  
b. Erythromycin  
c. Grapefruit Juice  
d. St. John's Wort  
e. Verapamil

**Correct answer:** D

INR (International Normalized Ratio) is a standardized way of reporting Prothrombin Time. Prothrombin Time (PT) is a measurement of the functioning of the Extrinsic Pathway of the coagulation cascade. (aPTT measures the intrinsic pathway, mnemonic: there are more letters in "aPTT" and more clotting factors in the intrinsic pathway). Warfarin therapy is monitored with INR or PT because it affects the Vit-K dependent clotting factors (II, VII, IX, X, C, S) and Factor VII specifically is part of the extrinsic pathway. Normal INR is between 0.9-1.3. This patient has an INR of 0.5, which means her blood is more likely to clot than normal (or is desirable for a person with a mechanical heart valve). Faster PT/INR, means clotting occurs more readily. This should not happen if Warfarin level in the blood are high.

The question is about liver metabolism of warfarin. Warfarin is metabolized by P450 enzymes in the liver. This is important because many other drugs either induce or inhibit the P450 isozymes, affecting warfarin levels. Inducers would speed up the metabolism of warfarin in the liver, and thus lower the serum levels (speeding up PT/INR). The only inducer listed is St. John's Wort (an over the counter herbal antidepressant). All the other drugs are inhibitors of P450 isozymes and would be expected to increase warfarin levels in the blood, and thus increase PT/INR.

2 "sing-song" mnemonics to help you memorize which drugs inhibit and which induce P450s is below:

Inhibitors: Sing this like a cheer ("2, 4, 6, 8 who do we appreciate..." ) that kind of melody....

"-Navir, -Azole  
Erytho- and Cipro-  
Inhibit! Inhibit! They all Inhibit!  
Cimitidine,  
Class Four And  
Grape-Fruit Juice.  
Inhibit! Inhibit! They all Inhibit!"

(The P450 inhibitors: All the HIV Protease inhibitors (end with suffix- navir), all the "azole" antifungals, Erythromycin (but none of the other macrolides), Ciprofloxacin and all the other fluoroquinolones, Cimitidine, Class IV Anti-arrrhymatics (use-dependent Ca channel blockers)= Diltiazem, Verapamil, and grapefruit juice)

And for the inducers: .. this is a rhyme, like a nursery rhyme...

"Treat TB and the mind,  
Induce the P450 enzyme"

Inducers of P450:  
TB drugs: Rifampin and Isoniazid  
"Mind" drugs (really barbituates, antiseizure drugs, and herbal antidepressants): Phenobarbital, Phenytoin, Carbamezapine, and St. John's Wort (herbal antidepressant)
A patient presents to the emergency room with a urinary tract infection. In reviewing your knowledge of antibiotics, you recall that which of the following is bacteriostatic versus bactericidal?

a. Aminoglycosides
b. Cephalosporins
c. Fluoroquinolones
d. Macrolides
e. Penicillin

**Correct Answer:** D

Macrolides are bacteriostatic agents that inhibit protein synthesis by binding to the 50S subunit of the bacterial ribosome. They are used against intracellular pathogens, including Mycoplasma and Legionella, and can also be used as alternatives in case of penicillin allergy. If used in high doses, they may be bactericidal.

Aminoglycosides (Choice A) are bactericidal agents that inhibit protein synthesis by binding to the 30S subunit of the bacterial ribosome. They are used against gram-negative aerobes, and can be used in conjunction with ampicillin for complicated urinary tract infections and certain forms of meningitis.

Cephalosporins (Choice B) are bactericidal agents that share a similar mechanism of action to penicillin. They act by inhibiting cross-linkage of the bacterial cell wall, compromising their structural and osmotic integrity. Cephalosporins are divided into four different classes based on their antimicrobial activity and beta-lactamase resistance.

Fluoroquinolones (Choice C) are bactericidal agents that inhibit bacterial DNA gyrase, thereby blocking replication of bacterial DNA. They are used primarily against gram-negative organisms. Coverage against gram-positive organisms is variable, although certain fluoroquinolones are commonly used to treat community-acquired pneumonia.

Penicillins (Choice E) are bactericidal agents that inhibit cross-linkage of the bacterial cell wall, compromising their structural and osmotic integrity. They are most effective against gram-positive organisms and are often used with aminoglycosides for a synergistic effect.
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- a. Aminoglycosides
- b. Cephalosporins
- c. Fluoroquinolones
- d. Macrolides
- e. Penicillins

**Correct answer: D**

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Penicillins (Choice E) are bactericidal agents that inhibit cross-linkage of the bacterial cell wall, compromising their structural and osmotic integrity. They are most effective against gram-positive organisms and are often used with aminoglycosides for a synergistic effect.
A 45 year old man own a small appliance store that he runs with his wife. His wife report that a week ago, he began talking about a new theory he had for making money in the state lottery. When his wife told him that he did not make any sense, he began screaming "I am a genius!, you don’t know what you’re talking about!, I am going to be filthy rich!” A few days later he secured a business loan and promptly used the money to purchase lottery tickets. "Nothing can stop me now” he told his wife. She said that he had slept little or late and spends his time working feverishly on lottery theory and rearranging merchandise at the store. Which of the following drugs would be most appropriate for long-term treatment of this patient?

a. Alprazolam  
b. Chlorpromazine  
c. Fluoxetine  
d. Lithium Carbonate  
e. Phenelzine

**Correct answer:** D

D. is correct, this patient is having a manic episode. Lithium is effective in decreasing manic behavior and in reducing the magnitude and frequency of mood swings. At the beginning of Lithium, neuroleptics or benzodiazepines may be given to control the patient, although they would do nothing for the underlying mood disorder. Antidepressant drugs may be required as adjuvants to lithium therapy to control depression.

A. Alprazolam is a benzodiazepine that might be used initially as a sedative/hypnotic.

B. Chlorpromazine is a phenothiazide antipsychotic that might be used initially to sedate the patient.

C. Fluoxetine is selective serotonin reuptake inhibitor (SSRI) antidepressant and E. Phenelzine a monoamine oxidase inhibitor (MAOI) antidepressant, might be used as adjuvants to lithium in this patient to control his depression, but would not address his mania.
A patient is diagnosed with gout and the doctor starts him on colchicine. This drug acts by which of the following methods and to treat which type of gout?

- a. Inhibits xanthine oxidase to decrease conversion of xanthine to uric acid; used to treat acute gout
- b. Inhibits xanthine oxidase to decrease conversion of xanthine to uric acid; used to treat chronic gout
- c. Inhibits the reabsorption of uric acid in the proximal convoluted tubule; used to treat chronic gout
- d. Inhibits the reabsorption of uric acid in the proximal convoluted tubule; used to treat acute gout
- e. Inhibits COX-2 to block the deposition of urate crystals in joints

correct answer: D

Colchicine blocks the reabsorption of uric acid in the proximal convoluted tubule, thereby limiting the amount of uric acid stores in the body available to deposit in joints. It is only used in the treatment of acute gout.
Patient experiences dyspnea on exertion...)

a. c
b. j
c. s
d. p
e. o

**Correct answer:** A
A 60 year-old male presents with a hot, red, swollen MTP joint of the big toe. Pt. complains that onset of pain was sudden and rates the pain as 7 out of 10. Past medical history is significant for high blood pressure, which is recently being treated with a thiazide diuretic. A joint aspiration is likely to reveal the following:

a. Bipyramidal shaped, positively refringent crystals  
b. Flat rhomboidal plates shaped  
c. Needle-shaped, negatively birefringent crystals  
d. Rhomboid shaped, positively birefringent crystals  
e. Spherically-shaped clumps

**correct answer:** C

The patient is experiencing a gout attack, which is commonly brought on by thiazide diuretics (which inhibit uric acid secretion). Gout crystals are needle-shaped and negatively birefringent under polarized light. The other answer choices describe pseudogout (rhomboid shaped, positively birefringent), osteoarthritis (spherical clumps), calcium oxalate stones (bipyramidal, positively refringent), and hypercholesteremia stones (flat rhomboidal plates).
A patient is diagnosed with gout and the doctor starts him on probenecid. This drug acts by which of the following methods and to treat which type of gout?

- a. Inhibits xanthine oxidase to decrease conversion of xanthine to uric acid; used to treat acute gout
- b. Inhibits xanthine oxidase to decrease conversion of xanthine to uric acid; used to treat chronic gout
- c. Inhibits the reabsorption of uric acid in the proximal convoluted tubule; used to treat chronic gout
- d. Inhibits the reabsorption of uric acid in the proximal convoluted tubule; used to treat acute gout
- e. Inhibits COX-2 to block the deposition of urate crystals in joints

**Correct answer: C**

Probenecid blocks the reabsorption of uric acid in the proximal convoluted tubule, thereby limiting the amount of uric acid stores in the body available to deposit in joints. It is only used in the treatment of chronic gout.

Answer choice B describes allopurinol. Allopurinol also used to prevent tumor lysis urate nephropathy.

Acute gout is treated by NSAIDs and if allergic to that - colchicine. Colchicine works by depolymerizing microtubules thus inhibition leukocyte movement and degranulation - notable GI side effects so 2nd line drug after NSAID like indomethacin.
A 65 year old man presents with auditory hallucinations saying he “hears the voice of god.” He also seems to talk without much expression or emotion. He claims that his neighbors are actually aliens who threaten to kidnap him. He has had these thoughts and behaviour for the last 2 years. In the ED, he is given an injection, but a few hours later his vitals are noted to be: BP 190/100; P 120; Temp 103; RR 21. A CPK is ordered and is found to be very elevated. What medication should be administered immediately?

a. Dantrolene Sodium  
b. Haloperidol  
c. Clozapine  
d. Benztropine  
e. Amantadine

_correct answer: A

This man’s psychosis, flat affect, bizarre delusions and behavior for at least 6 months are indicative of schizophrenia. He was treated with an anti-psychotic agent (such as haloperidol or risperidone) and subsequently developed neuroleptic malignant syndrome. NMS is a potentially life-threatening side effect of both typical and atypical neuroleptics. It is characterized by elevated vital signs, autonomic instability and a rising CPK. It is treated with dantrolene (Choice A) and/or a dopamine agonist.

Haloperidol (Choice B) is an anti-psychotic that causes NMS but is not used to treat it.

Clozapine (Choice C) is also an anti-psychotic but again may cause NMS.

Benztropine (Choice D) is an anti-cholinergic drug used to reverse the dystonic states caused by anti-psychotic agents. It is also used as a second-line treatment for Parkinson’s disease.

Amantadine (Choice E) is a drug used in Parkinson’s Disease.
A 22-year-old man is admitted to inpatient psychiatry after an attempt to strangle a stranger on the street in order to "squeeze the demons out" at the command of a voice in his head. He was put on a standing dose of haloperidol. Which of the following long-term adverse effects can be attributed to haloperidol?

a. tardive dyskinesia
b. Excess salivation
c. orthostatic hypotension
d. Nausea
e. Pruritus

**Correct answer:** A

A is correct: Haloperidol is a high-potency typical antipsychotic. Unlike low potency antipsychotics such as chlorpromazine, haloperidol has a much stronger adverse effect profile due to its high affinity for D2 receptors. Tardive dyskinesia is due to the strong dopamine receptor blockade. The blockade leads to long-term up-regulation of D2 receptors and the resultant hyperkinesia.

B is incorrect: Excess salivation would be due to stimulation of cholinergic receptors.

C, D, and E are incorrect: Antihistamine effects, in fact, include anti-nausea (antiemetic) and decreased itching (pruritus). Hypotension would be expected with alpha-1 blockade.
A 85-year-old male is brought to your office by his wife. She complains that his memory has been gradually getting worse and that he has difficulty buttoning his shirt. On interviewing him, you find that he has trouble finding words. Which drug is contraindicated for his condition?

a. Benztropine  
b. Donepezil  
c. Galantamine  
d. Memantine  
e. Rivastigmine

**Correct answer:** A

This patient has symptoms consistent with Alzheimer's Disease. He has memory impairment, apraxia, and difficulty finding words. Drugs given to delay progression of disease are cholinesterase inhibitors (donepezil, rivastigmine, galantamine) and NMDA antagonists (memantine).

Benztropine is a cholinoreceptor blocker; although Parkinson's disease is not well understood, it is clear that benztropine should not be used in conjunction with any of the cholinesterase inhibitors.
A patient in the psychiatry wards has been abusing a particular drug with a very low safety margin. This drug causes respiratory depression, slowness of movement and slurred speech. His withdrawal episodes have included rebound anxiety, seizures and wildly fluctuating hypertension.

This drug most likely belongs to which of the following classes?

a. Amphetamines  
b. Barbiturates  
c. Benzodiazepines  
d. Hallucinogens  
e. Opioids

**Correct answer: B**

Barbiturates (B) produce respiratory depression and slurred speech with the withdrawal effects described in the question stem. Cardiovascular collapse is also a potentially life threatening consequence of withdrawal. Notably, barbiturates have a very low safety margin.

Amphetamines (A) produce the opposite effects described, including agitation, tachycardia, euphoria, and prolonged attentiveness.

Benzodiazepines (C) would produce similar effects as barbiturates, but are widely used because they have a much greater therapeutic window.

(D) Hallucinogens and (E) Opioids possess a more complex abuse profile can themselves cause anxiety or seizures, respectively.
A 6-year-old child is brought to your office for evaluation of seizures. While examining the child, you hand her a pinwheel to blow on rapidly. After she blows on the pinwheel to make it spin very quickly, she suddenly stops, appears to stare of into the distance, and seems to "drop out". Within a minute, she is back to normal. EEG studies reveal generalized 2-3 Hz spike and wave discharges.

Which of the following drugs would you try as first line therapy for this child?

- a. Carbamazepine
- b. Ethosuximide
- c. Phenytoin
- d. Valproic acid
- e. Verapamil

**Correct answer:** B

This child has generalized seizures of the absence type (formerly called "petit mal"). These occur most often in children and can be induced by hyperventilation. (Asking the child to blow on the pinwheel rapidly simulates hyperventilation). The EEG findings described are classic for absense seizures.

Absence seizures are best treated by drugs that specifically target T-type Ca+ currents, which are the believed to be involved in providing the pacemaker current for thalamic neurons which then generate the generalized rhythmic discharge in the cortex during absence attacks. The only two drugs listed that target the T-type Ca current are (B) ethosuximide and (D) Valproic Acid. Ethosuximide has much better safety profile than valproic acid and thus is considered first line treatment for absence type seizure disorder in children.

The other two anti-seizure drugs (A) carbamazepine and (C) phenytoin are contra-indicated in absence seizures. Both are classified as "membrane stabilizer" drugs, and are most effective against partial seizures and generalized tonic clonic seizures (formerly called "grande mal").

Choice (E), verapamil is a L-type Ca+ blocker, a Class IV cardiac antiarrhythmic drug. It does not treat seizures.

**Sources:**
A five-year-old boy with a history of Tourette syndrome is brought to the pediatrician by his mother. She reports that, over the past six months or so, her son has started having difficulty in school: he easily loses focus, procrastinates, and shows poor impulse control. At the last parent-teacher conference, his teacher complained he is constantly "bouncing off the walls" and generally disrupting class.

If his pediatrician were to treat these new symptoms pharmacologically, which of the following drugs would be most appropriate?

a. Atomoxetine  
b. Dextroamphetamine  
c. Methylphenidate  
d. Modafinil  
e. Risperidone

**Correct answer:** A

Attention-deficit hyperactivity disorder (ADHD) is a developmental disorder characterized by academic problems (short attention span, poor concentration, etc.) and behavioral problems (impulsivity, hyperactivity, etc.). The academic problems arise from insufficient noradrenergic signaling in the brain as norepinephrine reduces noise and enhances executive function, whereas the behavioral problems are a result of overactive dopaminergic signalling.

Standard treatment of this condition involves using CNS stimulants, such as methylphenidate and dextroamphetamine, to bring about increased dopaminergic signaling in the brain.

However, using dopaminergic drugs in this patient would exacerbate his Tourette syndrome, which involves overactive dopaminergic signalling in the thalamus, basal ganglia, and frontal cortex. In this case, atomoxetine (a norepinephrine reuptake inhibitor) is best as it will improve the patient's concentration without worsening his tics.
40-years-old engineer driving a train got involved in a crash collision with an oncoming train. He has history of glaucoma. Tetrahydrocannabinol was found in his urine. The most likely responsible drug for this accident is?

- a. Pilocarpine
- b. Amphetamines
- c. Atropine
- d. Marijuana
- e. Cocaine

**Correct answer:** D

Marijuana is most common illegal drug of addiction used in US. It contains tetra-hydro-cannabinol (THC), a psychoactive stimulant, which binds to receptors in different areas in the brain, e.g., cerebellum and hippocampus. Its clinical application varies from lowering intraocular pressure in glaucoma patients, decreasing nausea and vomiting in patients receiving chemotherapy, to analgesia. Delayed reaction time is most serious in those patients who operate on vehicles.

Pilocarpine is used in acute angle glaucoma; however, atropine causes glaucoma (acute angle). Amphetamine can cause hallucination. Cocaine can cause midriasis, ventricular arrythmias and sudden cardiac death.
A 27 year old patient with trigeminal neuralgia has been admitted for dysarthria and ataxia. She has vertical and horizontal nystagmus. Attending in charge tell you there is no specific treatment for this type of toxicity. Which drug causes the nystagmus in this patient?

a. Tacrine
b. Nedocromil
c. Phenytoin
d. Cisapride
e. Candesartan

**Correct answer:** C

There is no cure for trigeminal neuralgia, but Phenytoin other anti-convulsants and even botox can be used. Is important to understand the physio-path of trigeminal neuralgia, since USMLE will hammer this, is one point you dont want to loose.

A) Ache inhibitor used for Alzheimers disease, is hepatotoxic and it causes GI Bleeding.
B) Inhibits mast cell degranulation by increasing the Cl channel efflux, to make the cell more negative. It can cause severe Pulmonary Eosinophilia.
D) Is used for gastroparesis, it is no longer used since it can cause torse de Pointe
E) it blocks angiotension II receptors, it can cause fatal renal toxicity.
What would be the most likely medication to cause vertical and horizontal nystagmus in a 27 year old patient treated for trigeminal neuralgia?

a. Tacrine
b. Nedocromil
c. Phenytoin
d. Cisapride
e. Candesartan

correct answer: C
While there is no cure for trigeminal neuralgia, phenytoin other anti-convulsants and botox can be used. Generally, anticonvulsants are used for many kinds of neuropathic pain. For USMLE Step 1, it is a good idea to associate drugs with 'classic' side effects, that is side effects that occur more often due to that drug than any other drug. This idea is especially useful when studying chemotherapeutic and hypoinsulinemics.

A) Tacrine is an acetylcholinesterase (AChE) inhibitor used in Alzheimer's disease. It classic side effects are hepatotoxicity and GI bleeding.
B) Nedocromil inhibits mast cell degranulation and is used in the treatment of asthma. It belongs to the same class of drugs as cromolyn sodium. It hyperpolarizes the mast cell membrane via increased Cl channel efflux, Its classic side effect is severe pulmonary eosinophilia.
D) Cisapride was used for gastroparesis. It is no longer used because it can cause torsades de pointes. Its mechanism of action is as a 5-HT4 agonist.
E) Candesartan is an angiotensin II receptor blocker (ARB) that can cause fatal renal toxicity. Easy way to remember is to see the ending letters - sartan (remember Losartan).
A 40 years old male is brought to the ER, with severe headache, blurred vision, hand tremor, blood pressure is 210/130 mmHg and heart rate 110 per min, after dining at a local pizza restaurant. He received four weeks of treatment for his impaired concentration, loss of energy, decreased sexual drive, rejection sensitive and increased sleep and appetite. The medication used by this patient was:

a. presynaptic selective serotonin uptake
b. presynaptic non selective monoamine uptake
c. non selective monoamine oxidase inhibitors
d. converts dopamine to 3-O-metyldopa (COMT) inhibitors
e. selective monoamine oxidase inhibitors

Correct answer: C

Non selective monoamine oxidase inhibitors such as phenelzine, trancylcypromine are not considered to be first line antidepressant medications because dietars restrictions (cheese, wine) producing a hypertensive crisis like this case, however are particulary useful in this patient with his atypical depression.

A) presynaptic selective serotonin uptake, SSRI antidepressants such as fluoxetine, sertraline, paroxetine that block re uptake of serotonin at the pre synaptic neuron.

B) presynaptic non selective monoamine uptake refers to tricyclic antidepressants such as imipramine, amitriptyline, clomipramine. TCA block norepinephrine or serotonin reuptake.

D) converts dopamine to 3-O-metyldopa (COMT) inhibitors such as talcapone and entacapone, enhance levodopa uptake and efficacy, used in parkinson disease.

E) selective monoamine oxidase inhibitors such as selegiline no tyramine interactions, initial treatment and adjunct to levodopa in parkinson disease.
A 48 year-old male comes in to your outpatient clinic complaining of severe anxiety and a throbbing headache. His vital signs show a blood pressure of 190/130 mmHg. His current medications include an anti-depressant. He says he is not under any extra stressful conditions, and his diet has been extremely healthy lately -- consisting primarily of soy, cheese, and avocados. What medication is he on?

a. Fluoxetine
b. Imipramine
c. Nortriptyline
d. Tranylcypromine
e. Venlafaxine

correct answer: D

The patient is on tranylcypromine, a monoamine oxidase (MAO) inhibitor. He is experiencing a hypertensive crisis, as a result of high levels of tyramine (found in cheese among other foods) ingestion. When ingested orally, MAOIs inhibit the catabolism of dietary amines. When foods containing tyramine are consumed, the individual may suffer from hypertensive crisis. If foods containing tryptophan are consumed, hyperserotonemia may result. The amount required to cause a reaction varies greatly from individual to individual, and depends on the degree of inhibition, which in turn depends on dosage and selectivity.
A 32 year old woman is admitted the hospital after complaining of urinary frequency, dysuria, and weakness. She is not sexually active and LMP was one week ago. She is afebrile, blood pressure is 118/80, and pulse is 68/min. Costovertebral angle tenderness is elicited. IV ceftriaxone is started. Two days later, the patient is afebrile and feels much better. Antibiotic susceptibility testing showed E. coli highly sensitive to ceftriaxone, gentamicin, ciprofloxacin, and trimethoprim/sulfamethoxazole (TMP/SMX). Which is the best step in the management of this patient?

a. Add ciprofloxacin to the regimen
b. Continue ceftriaxone
c. Discontinue antibiotic therapy
d. Switch to gentamicin
e. Switch to TMP/SMX

**Correct answer:** E

This patient has signs and symptoms suggestive of uncomplicated pyelonephritis, which responded well to parenteral antibiotic therapy. Patients with uncomplicated pyelonephritis can usually be switched to an oral antibiotic after 48-72 hours of parenteral therapy. The most reasonable step would be to switch to an antibiotic according to the susceptibility testing. TMP/SMX is a good choice, because it is relatively cheap, and she can continue on TMP/SMX for two weeks. There is no reason to add another antibiotic, or continue parenteral agents, which would require hospitalization.
A patient with newly diagnosed hypertension is treated with a medication that prevents concentration of the urine by abolishing the hypertonicity of the medulla. What area of the kidney is most likely to be affected as an adverse effect of the drug?

- a. Glomerulus
- b. Interstitium
- c. Collecting ducts
- d. Tubules
- e. Afferent arterioles

**Correct answer:** B

The medication described in the question is a loop diuretic, such as furosemide. In toxic levels, this drug can cause interstitial nephritis.

The collecting ducts and collecting tubules is the location of action for the K+-sparing diuretics and not a site of furosemide toxicity.

The action of loop diuretics occurs distal to the glomerulus and afferent arterioles and these areas are not affected by furosemide toxicity.
A 63-year-old man presents to the hospital with an extremely painful right big toe. He reports that the pain started last night and was unrelieved by pain medication. He denies any trauma to the toe. On exam, the right first metatarsophalangeal joint is red, swollen and extremely painful to palpation. Aspiration of the swollen joint reveals long, needle-shaped, negatively birefringent crystals under polarized light. Appropriate treatment is started, and the patient is told to avoid aspirin. For which of the following reasons is aspirin likely contraindicated in this patient?

a. Aspirin reduces renal clearance of uric acid
b. Aspirin does not provide analgesia
c. Aspirin mobilizes uric acid from body stores
d. Aspirin is inferior to nonsteroidal anti-inflammatory drugs for analgesia
e. Aspirin increases renal clearance of uric acid

**correct answer:** A

Aspirin is contraindicated in patients with gout because at low concentration, it can reduce renal clearance of uric acid and increase the hyperuricemia.

NSAIDS, such as indomethacin, are very effective for patients with gout for reducing inflammation and relieving pain, but this is not a reason why aspirin is contraindicated for patients with gout.

Aspirin would not provide adequate pain relief, but the reason why it is contraindicated is because of its effects on decreasing the clearance of uric acid.

Aspirin does not mobilize uric acid from body stores. This is the mechanism of action of uricosuric acids, such as probenecid.
A 35-year-old female complains of excessive urination during the past four days. She has a history of bipolar disorder for which she has been treated for the past 15 years. A water restriction test is performed, and no elevation in urine osmolality is observed following the administration of desmopressin. Which of the following most likely accounts for her recent onset polyuria?

a. Inhibition of cAMP synthesis in the renal tubule
b. Inhibition of inositol trisphosphate synthesis in the renal tubule
c. Stimulation of cAMP synthesis in the renal tubule
d. Stimulation of inositol trisphosphate synthesis in the renal tubule
e. Stimulation of aquaporin-2 channel insertion in the renal tubule

Correct answer: A

Polyuria in the context of long-term treatment of bipolar disorder suggests lithium toxicity. Lithium is the treatment of choice for bipolar disorder.

The clinical manifestations of lithium toxicity vary; they include neuromuscular excitability (including tremor, fasciculations, and rigidity), muscle weakness, ataxia, GI upset, bradycardia, and hypotension. Nephrogenic diabetes insipidus (DI) may develop in as many as 20% of patients receiving lithium chronically.

Nephrogenic DI is characterized by a defective renal response to ADH (antidiuretic hormone; also called vasopressin). ADH normally acts on cells of the renal collecting tubules. There it stimulates the vasopressin V$_2$ channel, a metabotropic channel coupled to a G$_s$ protein. G$_s$ stimulates adenylate cyclase, which synthesizes cAMP from ATP. The rise in intracellular cAMP triggers the insertion of aquaporin-2 channels into the plasma membrane, allowing the reabsorption of water.

A water restriction test is useful for discerning between central and nephrogenic DI. In central DI, the posterior pituitary gland does not produce sufficient ADH. In nephrogenic DI, there is inadequate response to ADH supplied by the pituitary gland. In the test, water is restricted for several hours until the patient's plasma osmolality reaches approximately 300 mOsm/kg. Demopressin, an ADH analog, is then administered and the patient's urine osmolality is subsequently measured. In central DI, desmopressin administration causes a rise in urine osmolality of 100% or more. In nephrogenic DI, there is usually less than a 15% rise in urine osmolality.

The exact mechanism by which lithium causes nephrogenic DI is not completely clear. It is thought that lithium accumulation in renal collecting tubule cells impairs the signaling between ADH and V$_2$ receptors. One hypothesis proposes that lithium prevents the rise in intracellular cAMP, while another suggests that lithium impairs signaling downstream of cAMP. Regardless of this controversy, choice A is the only one of the choices given that is compatible with the generation of nephrogenic DI.

Choice B and D: Inositol trisphosphate (IP$_3$) is a second messenger in G$_q$-coupled receptors. Of the receptors required to know for Step 1, only $\alpha_1$ (adrenergic), M$_1$ and M$_3$ (muscarinic), V$_1$ (ADH), and H$_1$ (histaminergic) receptors are coupled to G$_q$ proteins.

Choice C: Inhibition of V$_2$ receptors inhibits cAMP synthesis; it does not stimulate it.

Choice E: Inhibition of V$_2$ receptors decreases the insertion of aquaporin-2 channels in the renal collecting tubule.
A 57-year-old man with systolic heart failure comes to the Emergency Department with shortness of breath and swelling of his ankles. On physical exam, the patient is found to have 2+ pitting edema in both lower extremities. The patient is started on furosemide. In patients with systolic heart failure, medical therapy with furosemide is started in order to achieve which of the following?

a. Decrease in the risk of arrhythmia from electrolyte abnormalities
b. Improvement in mortality by decreasing the fluid load
c. Improvement in pedal edema with no change in amount of pulmonary edema
d. Improvement in symptoms by decreasing the fluid load
e. Increase in cardiac output by increasing diuresis

**correct answer:** D

The correct answer is improvement in symptoms by decreasing the fluid load (D). Furosemide inhibits sodium/potassium/chloride reabsorption in the ascending loop of Henle and distal renal tubule, which leads to increased excretion of water. Symptomatic patients with heart failure experience relief from pedal and pulmonary edema secondary to the decreased water load. Mortality is not affected however by this therapy (B). Furosemide therapy improves both pedal edema and pulmonary edema by decreasing the fluid load (C).

Furosemide therapy actually may increase the risk of arrhythmia from electrolyte abnormalities, especially hypokalemia (A).

If therapy with loop diuretics is too aggressive, cardiac output may decrease secondary to increased diuresis and hypovolemia (E). Physicians must maintain a delicate balance between symptom relief and adequate cardiac output.
An elderly Latino man comes to your clinic for a follow-up visit. He reports increased urination since last month, when you prescribed furosemide for his hypertension. He shows you his blood pressure diary, which shows a marked improvement in his blood pressure. However, he mentions that his heart feels like it is beating irregularly at times. What side effect of furosemide is probably causing this symptom?

a. Dehydration  
b. Hyperkalemia  
c. Hyponatremia  
d. Hypokalemia  
e. Hypomagnesemia

**correct answer**: D

Furosemide is a loop diuretic that inhibits the Na-K-2Cl co-transporter in the thick ascending loop of Henle in the kidney. It can cause dehydration and electrolyte depletion, particularly potassium. Signs of hypokalemia include cardiac arrhythmias, muscular weakness, muscle cramps, and constipation. More severe hypokalemia can lead to flaccid paralysis, hyporeflexia, and tetany. The hypokalemic effects of furosemide can be reversed with a potassium-sparing diuretic like spironolactone.
A 63-year-old man with a 5-year history of congestive heart failure comes to the emergency department because of a 1-month history of fatigue and labored breathing. Evaluation shows pulmonary edema. Furosemide is administered. Which of the following sets of physiologic changes is most likely following administration of the drug?

- a. Option A
- b. Option B
- c. Option C
- d. Option D
- e. Option E

**correct answer:** E
H is the correct answer.
A 42 year-old male is referred to your office because of asymptomatic hypertension. Currently, his blood pressure is 162/90 and 160/94 on two separate occasions. Two years ago he started on a regimen of hydrochlorothiazide, then metoprolol and amlodipine were added, yet his hypertension remains refractory to treatment. An arteriogram is shown. Which of the following drugs is contraindicated in this patient and for what reason? (The angiogram should show renal artery stenosis)

a. Simvastatin because it may transiently disrupt the integrity of the endothelial plaques
b. Hydralazine because it may result in rebound tachyrdia
c. Sodium nitroprusside because inhibiting a phosphodiesterase may lead to pulmonary hypotension
d. Enalapril because it may immediately lower GFR and lead to acute renal failure
e. Darbopoetin because it may induce polycythemia vera

correct answer: D
The answer is D. The patient has hypertension which is probably due to his bilateral renal artery stenosis as shown on the renal arteriogram. This results in decreased effective intra-arterial blood volume to the kidney and thus increased levels of renin secretion. Levels of angiotensin I, angiotensin II and aldosterone are all increased. Enalapril (or any ACE inhibitor or angiotensin receptor blocker like losartan) is contraindicated in bilateral renal artery stenosis because it may decrease GFR, decrease the already-low perfusion to the kidney and result in acute renal failure.
Choice A is incorrect; simvastatin is an HMG-CoA reductase inhibitor that would not be contraindicated in this patient and may be indicated based on his lipid profile.
Choice B is incorrect; hydralazine is an anti-hypertensive medication that works primarily at the arteries and arterioles to reduce blood pressure. It may be associated with tachycardia due to reduced blood pressure but would not be contraindicated here.
Choice C is incorrect; Sodium nitroprusside is a direct source of nitric oxide (it is not a phosphodiesterase inhibitor) and results in predominant venous dilation thus reducing cardiac preload, reducing cardiac demand and relieves the symptoms of angina.
Choice E is incorrect; darbopoetin is an erythropoetin analog used in treatment of anemia due to chronic renal disease, due to decreased erythropoietin production. It stimulates the bone marrow to increase red blood cell production.
A 48-year-old woman with a history of hypertension presents with headache and uncontrolled blood pressure. Medications include labetolol, ibuprofen and thiazide diuretics. Physical examination reveals a blood pressure of 190/115 mm Hg, heart rate of 98/min, respiration rate of 14/min, and decreased lower extremity pulses. Laboratory studies show a potassium level of 3.8 meq/L and a creatinine level of 1.7 mg/dL. The patient’s blood pressure is controlled with the addition of lisinopril and amlodipine. Laboratory findings on follow up visit were as follows:

- Blood urea nitrogen: 39 mg/dL
- Creatinine: 3.2 mg/dL
- Sodium: 143 meq/L
- Potassium: 5.3 meq/L
- Chloride: 98 meq/L
- Bicarbonate: 22 meq/L
- Urinalysis: Trace protein; hyaline casts/hpf

Which of the following is most likely responsible for these laboratory findings?

a. A Calcium channel blocker
b. Angiotensin converting enzyme inhibitor
c. Beta-adrenergic blocker
d. Distal tubule Na+/Cl⁻ channel blocker
e. COX pathway inhibitor

**Correct answer:** B

This question tests the ability to recognize patterns of renal failure and identifying possible causes such an abrupt change in renal function. In this vignette, it is clear that the patient has recently been placed on lisinopril and amlodipine to control her hypertension. A careful look at the labs reveals an elevated BUN, Creatinine and potassium levels. These strongly suggest ACE inhibitor (lisinopril) side effect on renal function. The blockage of ACE prevents the conversion of angiotensin I (ATI) to II (ATII). ATII functions to stimulate vasoconstriction of efferent arterioles, and stimulate aldosterone release that causes sodium reabsorption and potassium secretion.

Amlodipine a calcium channel blocker (choice A) is incorrect. Although Amlodipine was recently added to patients regimen to control HTN, it is not known to cause acute renal toxicity. Labetolol a beta-adrenergic blocker (choice C) is incorrect. Labetolol was part of the patients previous medicaltition and thus less likely to cause such an renal failure. Thiazide diuretic a distal tubule Na+/Cl⁻ channel blocker (choice D) is used to treat renal failure. Patient was already on a thiazide. Ibuprofen a cox pathway inhibitor (choice E) is incorrect. Although NSAIDs could cause an acute renal failure in this setting the rise in potassium is more consistent with an ACE inhibitor related renal toxicity.
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e. COX pathway inhibitor

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Thiazide diuretic, a distal tubule Na+/Cl- channel blocker (choice D) is used to treat renal failure. Patient was already on a thiazide.

Ibuprofen, a cox pathway inhibitor (choice E) is incorrect. Although NSAIDs could cause an acute renal failure in this setting the rise in potassium is more consistent with an ACE inhibitor related renal toxicity.
One month ago a 62-year-old man came to the emergency department for left shoulder pain and began therapy with ibuprofen; his other medications are low-dose aspirin, simvastatin, glipizide, and atenolol. His creatinine level is now 2.8 mg/dL; 7 weeks ago, it was 1.3 mg/dL. He is afebrile, heart rate is 57/min, respiration rate is 16/min, and blood pressure is 123/56 mm Hg. Urinalysis shows a specific gravity of 1.010, trace leukocyte esterase, 1+ protein, and trace blood. On microscopic examination there is moderate leukocyte casts/hpf. Urine sodium is 25 meq/L.

Which of the following is the most likely explanation for this presentation?

a. Simvastatin induced rhabdomyolysis with resultant renal toxicity
b. Atenolol induced hypotension resulting in acute renal hypoperfusion
c. Drug-induced acute renal failure
d. Goodpasture’s glomerulonephritis
e. Nephrotic syndrome

correct answer: C

Drug-induced acute renal failure from ibuprofen use is most likely the cause of his acute renal failure. NSAIDs have a side effect of causing interstitial nephritis. The presence of moderate leukocytosis, proteinuria and hematuria is consistent with NSAID related interstitial nephritis.

Simvastatin induced rhabdomyolysis with resultant renal toxicity (choice A) is incorrect. Rhabdomyolysis can cause renal failure, however symptoms will include muscle aches and the presence of moderate leukocyte casts indicates an infectious or interstitial process.

Atenolol induced hypotension resulting in acute renal hypoperfusion (choice B) is incorrect. Atenolol can cause bradycardia and hypotension but will least likely result in renal hypoperfusion.

Goodpasture’s glomerulonephritis (choice D) is incorrect. Goodpasture’s disease or anti-glomerular basement membrane disease is a rare condition characterised by rapid destruction of the kidneys and of the lungs. Although many diseases can present with these symptoms, Goodpasture’s syndrome is an autoimmune disease produced due to IgG antibodies produced against the basement membrane causing damage via a type II hypersensitivity reaction. Patients usually present with hemoptysis as well. Immunoflorescence studies staining for IgG deposits typically reveals the a linear pattern.

Nephrotic syndrome (choice E) is incorrect. Nephrotic syndrome is a nonspecific disorder in which the kidneys are damaged, causing them to leak large amounts of protein resulting in proteinuria at least 3.5 grams/day, hypoalbuminemia, hypercholesterolemia and increased clot formation. Patients with nephrotic syndrome do not present with hematuria and hypertension.
Which of the following is the most likely mechanism of the therapeutic effect of Bromocriptine in female patients with hyperprolactinemia to help increase fertility?

- a. Antiestrogen to decrease feedback inhibition of estrogen on pituitary gland and hypothalamus
- b. Dopamine receptor antagonist to increase prolactin release from the pituitary gland
- c. FSH and LH hormone replacement in female patients without a functional pituitary gland is not necessary
- d. Induction of FSH and LH release by direct action on the pituitary gland
- e. Dopamine receptor agonist to decrease prolactin release from the pituitary gland

**Correct answer:** E

The correct mechanism of action is dopamine receptor agonist to decrease prolactin release from the pituitary gland. In women with hyperprolactinemia, this therapeutic effect of decreasing prolactin secretion helps increase fertility. High levels of prolactin inhibit GnRH release and result in secondary amenorrhea.

Another way to remember the mechanism of action is to remember that dopamine is also known as Prolactin Inhibiting Factor. Thus, a Dopamine Agonist would act like Prolactin Inhibiting Factor and stop Prolactin release from the pituitary gland.

Bromocriptine is not a dopamine receptor antagonist.

A drug that acts as an antiestrogen to decrease feedback inhibition of estrogen on the pituitary gland and hypothalamus is clomiphene. The result of this drug's mechanism is to increase the release of FSH and LH to enhance ovulation.

Human menopausal gonadotropins and human chorionic gonadotropins act like FSH and LH to induce ovulation without a functioning pituitary.

GnRH preparations administered in a pulsatile manner induce FSH and LH release by acting on the pituitary gland and thus increasing fertility.
A 68y/o woman has breast cancer that is metastatic to the axillary lymphnodes. The primary tumor is excised and then chemotherapy, including Cyclophosphamide and Tamoxifen is started. Which of the following mechanisms best explains the likely beneficial effect on Tamoxifen on this patient?

a. Competition with cytoplasmic Estrogen receptors
b. Enhancement of cyclophosphamide cytotoxic therapy
c. Inhibition of residual Estrogen synthesis by the ovaries and Adrenal glands
d. Retardation of tumor growth by its androgenic activity
e. Stimulation of membrane-bound progestin receptors

**correct answer:** A
Tamoxifen works as a competitor on Cytoplasmic Estrogen receptors
A patient presents to your office complaining of thinning hair and a bald patch on the top of his head. Which of the following drugs (with its accompanying correct mechanism of action) is most appropriate to consider as therapy?

- a. ketoconazole: testosterone receptor blocker
- b. flutamide: 5-alpha reductase inhibitor
- c. flutamide: testosterone receptor blocker
- d. finasteride: 5-alpha reductase inhibitor
- e. finasteride: testosterone receptor blocker

**Correct answer: D**

Dihydrotestosterone (DHT) is essential for the development of male specific sex characteristics. It is formed from testosterone via the enzyme 5-alpha reductase. DHT plays a role in promoting male pattern baldness, exemplified by the description in the case above.

Finasteride is a 5-alpha reductase inhibitor that blocks the conversion of testosterone to DHT and can therefore help to promote hair growth by decreasing levels of DHT.
A 35 year old man confides to you that he has been having sexual problems that have been troubling him lately. On further questioning he reports that he noticed a reduction of his ejaculate compared to the past. Which of the following drugs would most likely explain his presentation?

a. Ketoconazole
b. Albuterol
c. Cocaine
d. Lithium
e. Furosemide

**Correct answer:** A

The antifungal ketoconazole is an azole that works principally by inhibition of cytochrome P450 14a-demethylase. This enzyme interferes with sterol formation of ergosterol, a component of fungal cell membranes. However, a side effect of ketoconazole can be inhibition of mammalian steroid biosynthesis. Interference of steroid production by the adrenals and the gonads reduces testosterone, causing decreased sperm volume (oligospermia).

The other medications listed are not known to cause oligospermia.
A 68 year old woman is diagnosed with mild hypertension and her physician elects to start treatment with a beta-blocker. The patient has a medical history significant for asthma for which she takes an albuterol inhaler as necessary, though she has not used it in three years. Which beta blocker is least likely to cause an exacerbation of her asthma?

- a. Nadolol
- b. Atenolol
- c. Pindolol
- d. Propranolol
- e. Sotalol

**Correct answer:** B

Atenolol is the only beta-1 selective beta-blocker. Other beta-1 selective beta-blockers include metoprolol, esmolol, and bisoprolol. All the other beta-blockers listed are nonselective (they block both beta-1 and beta-2).

In the lungs, the beta-2 receptor helps to mediate against bronchoconstriction (smooth muscle). This explains the use of beta-2 agonists such as albuterol and salmeterol in the treatment of reactive airway diseases such as asthma. Blocking this receptor may lead to a worsening of respiratory symptoms. Thus, most clinicians will recommend the use of a beta-1 selective agent when treating a patient with a history of lung disease (e.g. asthma, COPD, emphysema).
A 16-year-old boy with a history of asthma presents to the emergency room with shortness of breath for one hour unrelieved by home nebulizer treatments. He is able to respond to questions in one-word answers due to his difficulty breathing. On physical exam, his respiratory rate is 25 breaths per minute and he has bilateral wheezes. He is given supplemental oxygen, albuterol and ipratropium nebulizer treatments, methylprednisolone, and magnesium. After his third nebulizer treatment, his breathing becomes easier. His respiratory rate decreases to 16 breaths per minute, but his heart rate is now 116 beats per minute. Which of the following is most likely responsible for the increase in heart rate?

a. Albuterol
b. Ipratropium
c. Magnesium
d. Methylprednisolone
e. Oxygen

**Correct answer:** A

Albuterol is a beta-agonist that is the mainstay of treatment for acute asthma attacks. Although it acts mainly on beta-2-receptors to relax bronchial smooth muscle, it does have cross-reactivity and can cause tachycardia.

Ipratropium (Choice B) is often used in conjunction with a short-acting beta-agonist such as albuterol. It acts by blocking the action of acetylcholine at parasympathetic sites in bronchial smooth muscle, leading to bronchodilation. Tachycardia is not a commonly associated adverse effect.

Magnesium (Choice C) can be used in conjunction with standard emergency room asthma treatments to improve spirometric indices. Tachycardia is not a commonly associated adverse effect.

Methylprednisolone (Choice D) is a corticosteroid used as an anti-inflammatory agent in the treatment of asthma. Tachycardia is not a commonly associated adverse effect.

Oxygen (Choice E) is used in asthma treatment to counteract hypoxia. Tachycardia is not a commonly associated adverse effect.
A 46-year old man presented to his physician with an increasing cough productive of a yellowish sputum. In addition, he reports moderate shortness of breath on exertion. The man has a clinical history significant for substantial smoking in addition to an instance of sustained V-tach a year previous. Physical exam: patient with very shallow breathing and a barrel-shaped chest. Wheezing and distant heart sounds heard on auscultation. What is the mechanism of action of the drug most likely ordered for inhalational therapy for this man?

a. Blockade of adenosine receptors
b. Blockade of Nn and M3 receptors
c. Inhibition of phospholipase A2
d. Activation of beta-2 receptors
e. Blockade of H2 receptors

**Correct answer: B**
The correct answer is B. The drug ipratropium would work best here because of the contraindications to adrenergic stimulation because of the heart condition. Albuterol (choice D) and Theophylline (choice A) would not be good given the state of his heart, and steroids (choice C) are used more in the chronic treatment. Choice E is the MOA of Cimetidine, a drug used for GERD, H. Pylori, and other acid problems, not respiratory issues.
A 45-year-old male patient presents with excruciating pain in his right large toe. The pain woke him from sleep last night and continued throbbing throughout the night. The toe on examination is warm and tender. No other joints are affected. Patient admits to 3-4 alcoholic drinks per night and diet consisting mainly of steak and potatoes. What drug will be the best choice to treat this acute attack?

- a. Acetaminophen
- b. Allopurinol
- c. Indomethacin
- d. Morphine
- e. Probenecid

correct answer: C

The patient is suffering from gout. Gout often manifests in patients with a diet high in alcohol and red meat (high in purines). Most often the large toe is affected (podagra). Most commonly, the first line of treatment is a high dose of an NSAID such as indomethacin (option C). Gout is an inflammatory reaction to urate crystals in the joint and acetaminophen (option A) and morphine (option D) do not have anti-inflammatory properties. Allopurinol (A) and probenecid (E) are drugs used to treat chronic gout but are not useful in the acute setting since they might worsen the attack. They work as prophylaxis by decreasing uric acid levels.
A 20-year-old college student returns from a weekend camping trip. One week later, the patient develops an expanding annular rash with central clearing on his ankle and is being treated with doxycycline for 5 weeks. The drug acts by inhibiting which of the following processes?

a. Translocation of peptidyl molecule from A-site to P-site on mRNA
b. Formation of an active initiation complex by irreversible binding to 30S ribosomal subunit
c. Binding of the tRNA to the mRNA-ribosomal complex via the acceptor A site on the mRNA
d. Protein synthesis by binding to the 50S ribosomal subunit
e. Activity of the DNA gyrases

Correct answer: C
Doxycycline is a tetracycline. It is a bacteriostatic inhibitor of protein synthesis that acts by reversible binding to 30S ribosomal subunit to inhibit the acceptor side on the mRNA and thus block binding of tRNA to the mRNA-ribosomal complex. Doxycycline is used for the treatment of Stage 1 Lyme Disease.

The Aminoglycosides acts by irreversible binding to the 30S ribosomal subunit to inhibit formation of an active initiation complex. As a result, misreading of the mRNA template occurs.

Erythromycin is a macrolide that acts by reversible binding to the 50S ribosomal subunit of gram-positive microorganisms to inhibit translocation of the peptidyl molecule from A-site to P-site on mRNA.

Clindamycin and the Streptogranins are examples of drugs that bind to the 50S subunit to cause bacteriostatic inhibition of protein synthesis.

Drugs that work by inhibiting the activity of the DNA gyrase are the Quinolones. The result is a bactericidal effect. This class of drugs includes Ciprofloxacin and norfloxacin.
A 54-year-old man with newly diagnosed essential hypertension develops a painful right knee after taking his anti-hypertensive medication. Joint aspiration reveals needle shaped crystals that are negatively birefringent. What was the likely agent that precipitated his knee pain?

a. Clonidine  
b. Enalapril  
c. Hydrochlorothiazide  
d. Metoprolol  
e. Nifedipine

**Correct answer:** C

Hydrochlorothiazide (HCTZ) (choice C) is a common initial therapy for uncomplicated hypertension. It is a diuretic that inhibits the Na+/Cl- transporter in the early segment of the distal convoluted tubule. HCTZ is associated with hyperuricemia, along with hypercalcemia, hyperglycemia, and hyperlipidemia. Increased uric acid can precipitate an episode of gout, which this man experienced.

Clonidine (choice A) is an agonist of presynaptic alpha2-adrenoceptors on sympathetic neurons. Enalapril (choice B) is an ACE inhibitor. Metoprolol (choice D) is a beta1-adrenoceptor selective beta-blocker. Nifedipine (choice E) is a calcium channel blocker. All these choices are used as antihypertensives, but only HCTZ is associated with hyperuricemia.
A 65-year-old woman presents with a one month history of fever, malaise, and weight loss. She also complains of pain and stiffness in her shoulders and hips, and a unilateral headache with worsening vision. She has no history of trauma. What drug should she be treated with first?

a. Acetaminophen
b. Allopurinol
c. Ibuprofen
d. Infliximab
e. Prednisone

**Correct answer:** E

This woman has clinical signs suggesting polymyalgia rheumatica and temporal arteritis. The two diseases often present together. Although biopsy is needed to confirm temporal arteritis, prednisone (choice E) should be started because both diseases are steroid responsive, and prompt treatment is needed to prevent blindness due to occlusion of the ophthalmic artery.

Acetaminophen (choice A) and ibuprofen (choice C) are both cyclo-oxygenase inhibitors, and will relieve some of the muscle pain, but will not resolve the inflammation causing both of these conditions. Allopurinol (choice B) is a xanthine oxidase inhibitor used for the chronic treatment of gout. Infliximab (choice D) is a TNF-alpha antibody used to treat Crohn’s disease, rheumatoid arthritis, and ankylosing spondylitis.
One week after camping in the Adirondack Mountains, a 13-year-old boy presents to his doctor complaining of fever, nausea, headache, and the rash shown.

Of the given options, what is the preferred pharmacologic treatment?

- a. Azithromycin
- b. Aztreonam
- c. Ciprofloxacin
- d. Penicillin
- e. Tetracycline

**Correct answer:** E

This patient is suffering from Rocky Mountain Spotted Fever, as evidenced by the classic petechial rash of the arms and wrists including the palms and soles, with progression to generalized distribution. Also typical are fever, nausea, and headache. The causative agent is the obligate intracellular Gram-negative Rickettsia rickettsii. Tetracycline (choice E), a protein synthesis inhibitor, is the treatment of choice.

It's also important to remember that rashes on the palms and soles are characteristic of only a few conditions: RMSF, secondary syphilis, coxsackievirus (enterovirus), and adverse drug reactions.

Azithromycin (choice A) is a protein synthesis inhibitor, used for upper respiratory infections and sexually transmitted infections.

Aztreonam (choice B) is a powerful monobactam, used for Gram-negative rods like Klebsiella and Pseudomonas species.

Ciprofloxacin (choice C) is a fluoroquinolone used for Gram-negative rods infecting the urinary and GI tract.

Penicillin (choice D) is a beta-lactam, used for Gram-positive cocci & rods, as well as Gram-negative cocci and spirochetes.
A 25-year-old male presents complaining of several weeks of itching lesions on his legs and arms. On exam, there are sharply demarcated erythematous plaques with silvery scale on the lower legs and extensor surfaces of the arms bilaterally. Which of the following drugs would be most likely to alleviate the patient's symptoms?

a. Amoxicillin, oral
b. Betamethasone, topical
c. Fluconazole, oral
d. Ketoconazole, topical
e. Mupirocin, topical

**Correct answer:** B

This patient most likely has plaque psoriasis, an immune-mediated disease caused by hyperproliferation of keratinocytes in the epidermis and inflammatory cell infiltrates. While the details of psoriasis pathogenesis and treatment are complex, the mainstay of treatment for psoriasis are anti-inflammatory and immunomodulatory therapies (e.g.: topical steroids, topical calcineurin inhibitors, and anti-TNF-alpha agents). Of the answer choices, the only drug which falls under these categories is betamethasone (B), a moderately potent corticosteroid that can be administered topically.

Other answer choices (listed below) would not be effective in treating an immune-mediated condition such as psoriasis.

Amoxicillin (A) is an beta-lactam aminopenicillin antibiotic that is most effective against Gram-positive bacteria that do not produce beta-lactamase.

Fluconazole (C) is a triazole antifungal that inhibits cytochrome p450 and sterol C-14 alpha-demethylation. It is useful in treating a number of fungal infections, including candidiasis, cryptococcal meningitis, and tinea infections.

Ketoconazole (D) is an antifungal that inhibits fungal cell membrane ergosterol synthesis.

Mupirocin (E) is a topical antibiotic that selectively binds to bacterial isoleucyl transfer-RNA synthetase, inhibiting protein synthesis. It is used to treat bacterial skin infections.
A 50 year-old female with osteoporosis presents to the ED with severe back pain. She says that the pain began several days ago and was gradual in onset. She does not recall doing any heavy lifting or anything else out of the ordinary. A picture of her back is shown.

You decide to treat this patient with a drug that has which mechanism of action?

a. Inhibition of bacterial cell wall synthesis by binding and inhibiting a penicillin-binding protein and thus causing bacteriolysis.
b. Decreasing ergosterol synthesis and thus inhibiting cell membrane formation
c. Binding to DNA gyrase and topoisomerase IV thus inhibiting DNA synthesis
d. The drug is first converted by a virus thiamidine kinase then further phosphorylated into its active form within the cell, which is then incorporated into DNA causing chain termination.
e. The drug is a pyrophosphate analog which binds DNA polymerase ausing chain termination, but it does not require phosphorylation by a viral tyrosine kinase.

Correct answer: D
The answer is D. The patient has a case of herpes-zoster virus infection, or the shingles. This rash classically presents with extreme pain and is distributed along a dermatome, unilaterally. The drug of choice is acyclovir, which must be phosphorylated by a viral kinase to become active.
Choice A is incorrect; this answer choice describes the action of penicillins, which are commonly used for Gram positive bacterial infections.
Choice B is incorrect; this answer choice describes the mechanism of fluconazole, which is an antifungal agent.
Choice C is incorrect; this answer choice describes the mechanism of action of the fluoroquinolone antibiotics
Choice E is incorrect; this answer choice describes the mechanism of foscarnet which is a drug used commonly for cytomegalovirus retinitis as well as for acyclovir-resistant herpes viridae.

Ganciclovir is like acyclovir except that it can be activated by other viral kinase outside of HSV/VZV thymidine kinase but is more toxic than acyclovir (well tolerated).

Both Ganciclovir and foscarnet are nephrotoxic. Ganciclovir is also a bone marrow suppressor (luekopenia, neutropenia, thrombocytopenia)
A 19 year old male comes to your office with complaints of increasing shortness of breath while preparing for the upcoming basketball season. Upon further questioning, it is revealed that his father would suddenly lose and regain consciousness in his youth while participating in sports. On physical exam you perceive a systolic murmur on the left sternal border that is accompanied by an S4 gallop. His EKG is suggestive of left ventricular hypertrophy. Which of the following will most likely decrease the murmur that is heard upon auscultation?

- a. decreased preload, decreased afterload
- b. decreased preload, increased afterload
- c. increased preload, decreased afterload
- d. increased preload, increased afterload
- e. only decrease in afterload

correct answer: D

The patient in this case has hypertrophic obstructive cardiomyopathy (HOCM), which has an underlying pathology of septal hypertrophy and systolic anterior motion of the mitral valve, both resulting in left ventricular outflow obstruction. He presents with the most common complaint which is dyspnea secondary to decreased diastolic function of the heart (less common presentations of HOCM are syncope and sudden death). There can be a family history of HOCM, which has an autosomal dominant mode of inheritance. Systolic murmur on the left sternal border and an S4 gallop are characteristic findings for HOCM. Remember that an S4 gallop is indicative of a noncompliant stiff left ventricle and is usually found when there is left ventricular hypertrophy (which the patient has reflected on his EKG).

Because the underlying result of the pathology of HOCM is outflow obstruction, the murmur decreases (softens) when the left ventricular chamber size is larger and more full. This happens when there is an increased preload (i.e. squatting and leg raising) and an increased afterload (i.e. handgrip). {Choice D}

The murmur will increase (become louder) when there is a decrease in afterload (i.e. amyl nitrate) or if there is a decrease in preload (i.e. Valsalva or standing). {Choices A, B, C and E}
A 63-year-old alcoholic man presents to the Emergency Department with increasing abdominal girth and right upper quadrant pain. On physical exam, the patient has a narrow pulse pressure and an S3 and S4 are heard on auscultation of the heart. Examination of the right neck veins reveals venous pulsations that are best seen when the head of the bed is elevated to ninety degrees. The abdomen is distended with shifting dullness. Which of the following is the most likely cause of this patient’s ascites given the physical exam findings?

a. Alcoholic cardiomegaly given that the jugular venous pressure is elevated
b. Alcoholic cardiomegaly given that the jugular venous pressure is normal
c. Alcoholic cardiomegaly given that the patient has a narrow pulse pressure and gallop rhythm
d. Alcoholic cirrhosis given that there is shifting dullness on percussion of the abdomen
e. Alcoholic cirrhosis given that the jugular venous pressure is elevated

correct answer: A

This patient has ascites secondary to alcoholic cardiomegaly. This is differentiated from ascites from alcoholic cirrhosis by measuring the jugular venous pressure (JVP). An elevated JVP is seen with ascites and alcoholic cardiomegaly (B). In alcoholic cirrhosis, the JVP is often normal or low-normal, but not elevated (E). The narrow pulse pressure and gallop rhythm are consistent with alcoholic cardiomegaly, but do not help differentiate the etiology of the ascites (C). Shifting dullness on percussion of the abdomen is consistent with ascites, but does not reveal anything about the etiology (D).
A 25 year old man comes to your office for a routine physical exam for a new job. He has been in his usual state of health and has no new complaints. His vital signs are stable and his physical exam reveals no abnormalities. Part of his physical exam includes an EKG. Upon reading the EKG (pictured above), you notice an arrhythmia. Which type of block is most likely responsible for the strip pictured below?

a. Atrial flutter
b. First Degree atrioventricular (AV) block
c. Second Degree AV block – Mobitz I
d. Second Degree AV block – Mobitz II
e. Third degree AV block

**Correct answer:** B

The picture is that of first degree AV block (choice B). It is defined as a prolongation of the PR interval of greater than 200 msec, or one large box on an EKG strip. The PR interval represents the conduction delay through the AV node. A first degree AV block is usually asymptomatic.

Second degree block can be either Mobitz I (choice C) or Mobitz II (choice D). Mobitz I is when there is a progressive lengthening of the PR interval until a beat is dropped. This is represented by a P wave not followed by a QRS complex. This is also usually asymptomatic. Mobitz II are randomly dropped beats manifested by abrupt, nonconducted P waves.

Third degree AV block (choice E) is when the atria and ventricles are beating independently of each other. This is usually treated with a pacemaker.

Atrial flutter (choice A) is a rapid succession of identical atrial depolarization waves. It is usually characterized by a sawtooth appearance on EKG.
During an experiment, a 25-year-old man receives an intravenous infusion of angiotensin II at a rate that increases plasma concentrations approximately threefold. Which of the following sets of physiologic changes is most likely in this man?

- a. Increased renin, increased filtration fraction, increased efferent arteriole resistance
- b. Increased renin, increased filtration fraction, decreased efferent arteriole resistance
- c. Increased renin, decreased filtration fraction, increased efferent arteriole resistance
- d. Increased renin, decreased filtration fraction, decreased efferent arteriole resistance
- e. Decreased renin, increased filtration fraction, increased efferent arteriole resistance

**correct answer**: E
basic science: Physiology

question: 821

clinical science: Cardiovascular

The accompanying image is a synchronous tracing of aortic pressure, left atrial pressure, left ventricular pressure, left ventricular volume, and EKG throughout the cardiac cycle. Which of the following is the best description of events that occur at the beginning and end of Time Period 2?

a. Aortic valve closes, mitral valve closes
b. Aortic valve closes, mitral valve opens
c. Aortic valve opens, mitral valve closes
d. Diastolic inflow of blood from left atrium to left ventricle
e. Systolic ejection of blood from left ventricle to aorta

Correct answer: B

Aortic valve closes, mitral valve opens (Choice B) is the correct answer. At the beginning of time period 2, the aortic pressure tracing clearly shows the dichrotic notch associated with aortic valve closing. Near the end of Time Period 2, ventricular pressure drops below atrial pressure; accordingly, this is when the mitral valve opens.

Aortic valve closes, mitral valve closes (Choice A) is incorrect. Given the preceding rise in ventricular pressure, Time Period 2 is clearly at the end of systole. Although this period is when the aortic valve closes, the mitral valve has been closed during systole to prevent regurgitation into the atrium, and it will open when ventricular pressure drops below atrial pressure, allowing diastole.

Aortic valve opens, mitral valve closes (Choice C) is incorrect. Given the preceding rise in ventricular pressure, Time Period 2 is clearly at the end of systole. The aortic valve is open during systole, and closes to prevent regurgitation into the left ventricle; thus, it will be closing during Time Period 2. Additionally, the mitral valve has been closed during systole to prevent regurgitation into the atrium, and it will open when ventricular pressure drops below atrial pressure, allowing diastole.

Diastolic inflow of blood from left atrium to left ventricle (Choice D) is incorrect; as shown by the rise in the ventricular volume tracing, this event occurs during the time between Time Period 2 and Time Period 3 on the graph.

Systolic ejection of blood from left ventricle to aorta (Choice E) is incorrect. This event occurs between Time Period 1 and Time Period 2, as evidenced by the rise in ventricular pressure, and fall in ventricular volume.

Adapted from http://commons.wikimedia.org/wiki/File:Cardiac_Cycle_Left_Ventricle.PNG
Your team is puzzled by a murmur found on cardiac examination of a 30 year old patient. You notice that it occurs during systole. When you ask the patient to stand up, the murmur increases in intensity. The Valsalva maneuver also increases the intensity of the murmur. When the patient is supine, the murmur is decreased. Auscultation of the carotid arteries is unremarkable. His PCW pressure is within normal limits. What is most likely abnormality present in this patient?

a. aortic valve stenosis
b. aortic regurgitation
c. mitral regurgitation
d. dilated cardiomyopathy
e. hypertrophic cardiomyopathy

**Correct answer:** E

This patient most likely has hypertrophic cardiomyopathy, a condition that commonly leads to sudden cardiac death in young athletes. Dynamic outflow obstruction, which is usually present in HCM, is typically due to systolic anterior motion of the anterior leaflet of the mitral valve. Moreover, septal hypertrophy contributes to the obstruction of outflow of blood, which causes a systolic murmur. Decrease in preload reduces the diameter of the outflow tract, hence increasing the obstruction and the intensity the murmur. Standing and the Valsalva maneuver reduce preload by decreasing venous return.

A is incorrect. The murmur of aortic valve stenosis radiates to the carotids, and its intensity is decreased by a low preload.

B is incorrect. Aortic regurgitation occurs on diastole, hence incorrect, given that the patient murmur occurs on systole.

C is incorrect. Mitral regurgitation is a systolic murmur, but it increases the pulmonary capillary wedge pressure, as it increase pressure in the left atrium, which is transmitted to the pulmonary vasculature and increases the PCWP.

D is incorrect. Dilated cardiomyopathy can cause increased diameter of the heart valves, leading to valvular insufficiency. However these abnormalities tend to occur in later in the course of dilated cardiomyopathy. Additionally, mitral and tricuspid insufficiency would occur in the context of elevated PCWP, and aortic and pulmonic insufficiency occur on systole.
A 70 year old man was brought in the hospital by ambulance subsequent to chest pain that lasted 40 minutes. In order to assess the degree of stenosis of his coronary arteries, he underwent coronary angiography, a procedure during which a contrast dye was injected in his coronary vessels. To which phase of the ventricular action potential does visualization of the stenotic lesion correspond?

a. Phase 0  
b. Phase 1  
c. Phase 2  
d. Phase 3  
e. Phase 4

**Correct answer:** E

This question essentially tests the relation of the electrical activity of the heart to its mechanical function. The coronary arteries fill up during diastole, as the ventricles relax. Therefore a stenotic lesion in a coronary artery will be best visualized on diastole, because during that period the dye will fill the coronary circulation. Relaxation of the ventricle corresponds to Phase 4, the resting membrane potential, during which cardiac myocytes are highly permeable to potassium. The cell remains in this phase until it is stimulated by an electrical stimulus, causing it to depolarize again.

A is incorrect. Phase 0 is the upstroke of the action potential, corresponding to rapid depolarization of myocytes. Voltage-gated sodium channels open at that time. Shortly after depolarization, the ventricle starts to contract.

B is incorrect. Phase 1 of the action potential occurs with the inactivation of the fast Na+ channels. Voltage-gated K+ channels begin to open.

C is incorrect. Phase 2 is the plateau phase, sustained by a balance between inward movement of calcium through L-type calcium channels and outward movement of potassium.

D is incorrect. Phase 3 is the "rapid repolarization" phase, corresponding to closure of Ca2+ channels, and massive efflux of K+ through voltage-gated K+ channels that are still open. This is associated with the T wave not resting.
A 67 year-old man is evaluated after being treated for a broken arm. He sustained a small hairline fracture of the distal ulna after hitting his arm on a railing. He says it was only a minor bump and is surprised it caused a fracture. Serum chemistry is remarkable for elevated calcium at 11.6, low phosphate at 2.1, and slightly elevated alkaline phosphatase at 98. What is the most likely diagnosis?

a. Hyperparathyroidism
b. Osteoporosis
c. Paget's disease of bone
d. Renal insufficiency
e. Vitamin D deficiency

**Correct answer:** A

Elevated calcium, low phosphate, and elevated alkaline phosphatase are consistent with hyperparathyroidism, which can cause osteopenia, making this patient more susceptible to fracture after trauma. Paget's disease of bone would be expected to have a markedly elevated alkaline phosphatase (a byproduct of osteoblast activity). In vitamin D deficiency, low serum calcium would be seen. Renal insufficiency would lead to low serum calcium and elevated phosphate. Osteoporosis would not lead to electrolyte abnormalities.
A 36 year old patient is brought in complaining of palpitations. Upon physical exam, you notice an irregular rate. He says he feels hot, anxious, and is tremulous. Thyroid studies yield elevated thyroid hormones. TBG is below normal. Examination of the neck is unremarkable. Which of the following is likely to be found in the patient?

- a. A 20 year history of alcohol abuse
- b. Exposure of the neck to radiation 20 years ago
- c. A family history of heart disease
- d. A personal history of Parkinson disease
- e. None of the above

**Correct answer:** A

Low levels of TBG are most likely secondary to liver disease. A chronic history of alcoholism is one of the most common causes of hepatic cirrhosis, which leads to reduced production of proteins, such as TBG.

B is incorrect. Exposure to radiation to the neck can lead to thyroid cancer, but the case does not provide information that makes us suspect cancer. Also the normal neck exam argues against a thyroid malignancy.

C is incorrect. Although family history heart disease may be implicated in a young man with atrial fibrillation, the full picture of the case argues for hyperthyroid state (tremor, high body temperature, anxiety). Also, heart disease would not affect TBG levels.

D is incorrect. Parkinson disease causes tremor, but does not account for the other manifestations of the patient’s condition.
Destruction of the supraoptic nuclei in the hypothalamus would cause which of the following conditions?

- a. Anorexia
- b. Central Diabetes Insipidus
- c. Inability to Breastfeed
- d. Obesity
- e. Sleep disturbances

**Correct answer:** B

B. The supraoptic nuclei are the site of ADH production. ADH then travels down axons to the posterior pituitary where it is secreted in response to increased serum osmolarity or low blood volume. Lack of ADH is the cause of central diabetes insipidus.

A. Damage to the lateral area would cause reduced food intake
C. Damage to the paraventricular nuclei would cause decreased oxytocin production and breastfeeding difficulties.
D. Damage to the ventromedial nucleus would cause obesity
E. Damage to the suprachiasmatic nuclei would disrupt circadian rhythm and sleep patterns
Which of the following hormone's has the most potent stimulator effect on pancreatic secretion?

a. Secretin  
b. CCK  
c. Histamine  
d. Serotonin  
e. Insulin  

**Correct answer:** A

Secretin is secreted by the mucosa of the duodenum and jejunum in response to an acidic chyme. Secretin stimulates the secretion of large quantities of sodium bicarbonate by the ductal epithelium of pancreas. Without fluid secretion most of the enzymes remain temporarily stored in the acini and ducts until fluid is secreted.
after food which hormone acts on the stomach and which hormone increases or decreases as shown in graph. Pictures are shown in increasing and decreasing graph representation and student has to click right answer

- a. secretin
- b. gastrin
- c. somatostatin
- d. insulin
- e. CCK

Correct answer: E

After fatty meal CCK is released for fatty meal digestion
**basic science: Physiology  question: 62  clinical science: Hematology**

**Erythrocytes obtain their energy primarily from which process?**

a. Glycolysis  
b. Lipid metabolism  
c. Oxidative phosphorylation  
d. Photosynthesis  
e. Pyruvate metabolism

**correct answer:** A

Erythrocytes do not have any membrane-bound organelles (such as mitochondria). Therefore, they can only obtain their energy through glycolysis.
A 4 year old boy presents with Acute Lymphoblastic Leukemia. Laboratory tests show that the Terminal deoxynucleotidyl transferase (TdT) is positive. What is the biological function of TdT?

- a. Class Switch Recombination
- b. Antigenic Diversity
- c. Heavy Chain Expression
- d. Apoptosis
- e. RNA splicing

correct answer: B

TdT edits the sequences of V, D, and J exons and thus helps to create a diverse repertoire of immunoglobulins.
A 23-year-old man who is planning to climb Mount Kilimanjaro comes to your office and asks you how to prevent altitude sickness. He knows that gradual ascent and prevention of dehydration are important, but wants some pharmacologic help to make sure he does not get sick. You tell him that acetazolamide, a bicarbonate-wasting diuretic, may be useful. What is the mechanism by which acetazolamide decreases tissue hypoxia in a low-oxygen environment?

a. Decreasing oxygen utilization by tissues
b. Hyperventilation
c. Increasing cardiac output
d. Increasing oxygen-carrying capacity by increasing hematocrit
e. Vasodilation and better perfusion of vital tissues

Correct answer: B

The question stem tells you that acetazolamide is a bicarbonate-wasting diuretic. This means that bicarbonate is lost in the urine. This results in a metabolic acidosis. One of the physiologic mechanisms of compensating for a metabolic acidosis is hyperventilation (in order "to blow off CO2"). Peripheral chemoreceptors in the carotid and aortic bodies detect the increase in pCO2, and respiration is stimulated, allowing for better ventilation and increased oxygen saturation.
A baby is born tachypneic and cyanotic with SaO2 80% in the upper extremities and 99% in the lower extremities. The hypoxemia does not respond to 100% oxygen and a congenital heart defect is suspected. Emergent cardiac catheterization reveals transposition of the great vessels. Which of the following can be given to the baby to maintain a patent ductus arteriosus until surgical correction can be performed?

a. PGD2  
b. PGE2  
c. PGE1  
d. PGF2  
e. PGI2

**Correct answer:** B

Prostaglandins are formed through the arachidonic acid cascade in response to several stimuli, including inflammatory mediators. Each prostaglandin has several, and often antagonistic effects. PGE1 maintains a patent ductus arteriosus (PDA) even in a normal neonate for up to a week. In the case of transposition of the great vessels, this communication between pulmonary and arterial circulation is crucial to rescue the abnormally separated pulmonary and systemic circulations. During fetal development, a PDA is maintained to bypass the high resistance, non-respirating pulmonary circulation. This is one of the reasons NSAIDs, which block prostaglandin production, are contraindicated during pregnancy.

PGD2 causes bronchodilation, platelet aggregation, and vasodilation. PGI2 (prostacyclin) has similar effects as PGD2, with the addition of hyperalgesia and mucus secretion in the stomach. PGF2 causes bronchoconstriction and myometrial contraction.

Main effects of PGE2 include increased renal blood flow and diuresis but is not responsible for the patent PDA.
A 75-year-old man has had increasing shortness of breath with exertion during the past 2 weeks. He has a 25-year history of hypertension well controlled with diuretics. Two months ago, serum urea nitrogen and creatinine concentrations were within the reference ranges. His pulse is 98/min, respirations are 19/min, and blood pressure is 180/100 mm Hg. The lungs are dull to percussion at the bases, and crackles are heard one-third of the way up bilaterally. Cardiac examination shows increased jugular venous pressure, an S3 gallop, and no murmur. There is 3+ pitting edema of the lower extremities. Serum studies are above. The patient most likely has which one of the following types of acid-base disturbance?

a. Metabolic acidosis
b. Metabolic alkalosis
c. Respiratory acidosis
d. Respiratory alkalosis
e. No acid-base disturbance

correct answer: A

Please revise this explanation
A 72 year old man has a cardiac arrest and despite resuscitation efforts, dies 10 minutes later. When he is examined at the morgue, his muscles are frozen in a contracted state. The underlying physiologic reason for this is:

a. Because there is no source of ATP, the myosin head cannot detach from actin and they remain bound together
b. Because there is no source of calcium, there can be no actin myosin cycling
c. Because there is no source of ATP, the actin cannot detach from tropomyosin and the filament remains frozen in position
d. Because there is no source of ADP and Phosphorus, there is no power stroke of the myosin head to move the muscle
e. Because there is an ATP pool left in the muscle, the actin-myosin complex remains active and continually contracted

**Correct answer: A**
This is the condition of "rigor mortis." When no ATP is present, the myosin head cannot detach from the actin filament and the muscle is locked in place.
A young man is running his first marathon. Which of the following actions is taking place in his skeletal muscle when he runs, relative to when he is at rest?

- a. Decreased CO2 production
- b. Decreased O2 demand
- c. Decreased metabolites concentration
- d. Increased arteriolar resistance
- e. Increased lactate formation

**Correct answer:** E

Skeletal muscle circulation is controlled by local metabolic factors during exercise. The increased oxygen demand also results in increased CO2 production. This results in lactate formation (correct answer), which causes arterioles to dilate, resistance to drop, and metabolite concentration to increase.
A 72 year old man has a cardiac arrest and despite resuscitation efforts, dies 10 minutes later. When he is examined at the morgue, his muscles are frozen in a contracted state. The underlying physiologic reason for this is:

a. Because there is no source of ATP, the myosin head cannot detach from actin and they remain bound together
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c. Because there is no source of ATP, the actin cannot detach from tropomyosin and the filament remains frozen in position
d. Because there is no source of ADP and Phosphorus, there is no power stroke of the myosin head to move the muscle
e. Because there is an ATP pool left in the muscle, the actin-myosin complex remains active and continually contracted

**Correct answer:** A

This is the condition of "rigor mortis." When no ATP is present, the myosin head cannot detach from the actin filament. When they remain locked together, the muscle remains locked in place. See the following diagram to better visualize the setup:

http://faculty.ircc.edu/FACULTY/TFischer/AP1/cross%20bridge%20cycle.jpg

The lack of ATP, not the lack of calcium availability (choice B), is the reason for debility of actin-myosin cycling.

It is the interaction of actin and myosin, not the interaction of actin and tropomyosin (choice C), that is responsible for rigor mortis.

Lack of ATP, not the lack of ADP and phosphorus (choice D), is responsible for rigor mortis.

Lack of ATP, not abundance of ATP (choice E), is responsible for rigor mortis.
A man falls asleep with his arm draped over a chair, and in the morning is unable to extend his wrist. The sensory deficit expected is:

a. The palmar and dorsal aspects of the little finger and ½ of the ring finger
b. Palmar aspect and dorsal tips of the index and middle fingers
c. The posterior arm and dorsal hand, not including the fingertips, little finger, ring finger, or ½ of the middle finger
d. The entire posterior arm
e. The entire hand

Correct answer: C

Know "Saturday night palsy"! The radial nerve is the "great extensor" and know its sensory distribution! (as well as those of the other nerves of the brachial plexus)
All are increased in case of isometric exercise except

a. heart rate
b. cardiac output
c. mean arterial pressure
d. temperature
e. systemic vascular resistance

**Correct answer:** E

The relative increase in cardiac output and mean arterial pressure is such that systemic vascular resistance unchanged
Cerebrospinal fluid (CSF) is produced by the choroid plexus epithelium which is a component of the blood-brain barrier. CSF can be sampled directly by lumbar puncture. CSF fluid and blood serum contain many of the same molecules but concentrations vary between the two. Which of the following molecules are found at higher concentrations in the CSF as compared to blood?

- a. Cholesterol
- b. Creatinine
- c. Glucose
- d. Potassium
- e. Protein

**correct answer:** B

Creatinine (choice B), magnesium, and chloride are found at higher concentrations in the CSF.

Glucose (choice C), cholesterol (choice A), protein (choice E), potassium (choice D), and calcium are found at lower concentrations in the CSF. Protein and cholesterol are generally excluded from the CSF because of their large molecular size.

Sodium is found at an equal concentration with serum.
Sound waves are quantified based on frequency and intensity. Intensity is measured in decibels (dB) which is based on a log scale.

\[ dB = 20 \log \left( \frac{P}{P_0} \right) \]

\( \left( \frac{P}{P_0} \right) \) is a power ratio where \( P \) represents sound pressure being measured and \( P_0 \) represents reference pressure measured at the threshold frequency. If the dB were to triple from 20dB to 60dB, what will be the change in the power ratio?

a. 10 times larger
b. 10 times smaller
c. 100 times larger
d. 100 times smaller
e. 1,000 times larger

**Correct answer:** C

The change in dB is 40dB, which corresponds to a doubling of \( \log \left( \frac{P}{P_0} \right) \). In order for \( \log \left( \frac{P}{P_0} \right) \) to double, the power ratio \( \left( \frac{P}{P_0} \right) \) must increase by a factor of 100 (choice C).

100 times smaller (choice D) would be the appropriate decrease of the power ratio if the dB decreased by 40dB.

10 times smaller (choice B) would be the appropriate decrease of the power ratio if the dB decreased by 20dB.

10 times larger (choice A) would be the appropriate increase of the power ratio if the dB increased by 20dB.

1,000 times larger (choice E) would be the appropriate increase of the power ratio if the dB increased by 60dB.
A 72 year old woman presents to her physician with the complaint of loss of vision. Visual field examination reveals anopia of her right upper quadrant visual fields bilaterally. What is the most likely location of the lesion responsible for this defect?

a. Left optic nerve  
b. Left parietal lobe  
c. Left temporal lobe  
d. Right optic tract  
e. Right parietal lobe

**Correct answer:** C

This woman has a right upper quadrant anopia caused by a lesion of her left temporal lobe. Meyer's loop runs in this lobe and contains the inferior optic radiations, which contain fibers supplying the upper visual fields of the opposite side. Recall that the optic chiasm contains crossing fibers which represent bilateral temporal vision. Fibers representing nasal vision remain on the same side as they extend from the eye back into the brain. Therefore, the right optic tract will contain fibers representing the right nasal visual field and the left temporal visual field. The optic tract leads to the lateral geniculate body, which divides into the inferior optic radiations (Meyer's loop in the temporal lobe) and the dorsal optic radiations (in the parietal lobe). While the temporal lobe loop carries fibers representing the superior visual field, the parietal lobe loop carries fibers representing the inferior visual field. Please refer to this diagram to help you reason:

http://faculty.washington.edu/chudler/vispath.gif

The left optic nerve (choice A) only contains fibers from the left eye. A lesion here would cause full loss of vision in the left eye.

The left parietal lobe (choice B) carries fibers representing the inferior visual field of the opposite side. A lesion here would produce a right lower quadrant anopia.

As noted above, the right optic tract (choice D) contains fibers representing the right nasal visual field and the left temporal visual field. A lesion here would produce a left homonymous hemianopia.

The right parietal lobe (choice E) carries fibers representing the inferior visual field of the opposite side. A lesion here would produce a left lower quadrant anopia.
A young boy puts a ring on his finger and soon forgets it is there. Which sensory receptor was the first to stop responding to the stimulus?

- a. Free Nerve Ending - C fibers
- b. Free Nerve Ending - A Delta fiber
- c. Meissner’s Corpuscle
- d. Pacinian Corpuscle
- e. Merkel’s Disks

**Correct answer:** C
Free nerve endings - pain and temperature sensation
Meissner’s Corpuscle - Position sense, fine touch. Because this receptor is rapidly adapting, it is the correct answer.
Pacinian Corpuscle - Vibration, pressure
Merkel’s Disk - Position sense, static touch, and slowly adapting.
A 15-year-old female presents to her pediatrician complaining of blood in her urine. She is otherwise healthy, with the exception of a sore throat approximately 2 weeks before. Review of systems is notable only for swollen ankles, which she denies having experienced before. On physical exam, her blood pressure is 145/85 and she has 1+ lower leg edema bilaterally. What serum study is most likely to be elevated?

a. Amyloid  
b. Anti-glomerular basement membrane  
c. Anti-streptolysin O  
d. Complement  
e. Immunoglobulin A

**correct answer:** C

The most common cause of nephritis syndrome is poststreptococcal glomerulonephritis. It develops approximately 10 to 14 days after a group A beta-hemolytic streptococcal infection, most commonly of the upper respiratory tract but also of the skin. Features of poststreptococcal glomerulonephritis include hematuria, edema, and hypertension. Serum levels of anti-streptolysin O may be elevated. The condition is generally self-limited and resolves in a matter of weeks to months. Rare cases may develop into rapidly progressive glomerulonephritis.

Elevated levels of amyloid (Choice A) are not found in poststreptococcal glomerulonephritis. Acute phase serum amyloid A proteins may be elevated in several chronic inflammatory diseases including amyloidosis, atherosclerosis, and rheumatoid arthritis. Anti-glomerular basement membrane (Choice B) is found in Goodpasture’s syndrome, a form of rapidly progressive glomerulonephritis.

Complement (Choice D) levels are usually decreased in systemic lupus erythematous, a common cause of glomerular disease. Immunoglobulin A nephropathy, or Berger’s disease, is the most common cause of glomerulonephritis worldwide. Asymptomatic, recurrent hematuria may occur following an upper respiratory infection or exercise. IgA levels may be elevated.
Glomerular Filtration Rate (GFR) in a 27 year old patient is found to be 30 ml/min. Renal blood flow (RBF) is determined to be 1.2 L/min. Hematocrit (Hct) is currently 0.25. Six months ago, the patient’s Hct was 0.5. Assuming that GFR remains unchanged, which of the following is most likely true regarding the effect of this decrease in hematocrit on renal function?

a. Increased Renal Blood Flow
b. Increased Renal Plasma Flow
c. Increased Filtration Fraction
d. Unchanged Filtration Fraction
e. None of the above

**Correct answer:** B

This question asks you to have an understanding of the relationships between Renal Blood Flow (RBF), Renal Plasma Flow (RPF), Hematocrit (Hct), and Glomerular Filtration Rate (GFR). \[ \text{RBF} = \frac{\text{RPF}}{1 - \text{Hct}} \] This equation suggests that a decrease in Hct would lead to an increase in RPF.

Using the same equation, a decrease in Hct would cause a decrease in RBF, therefore (A) is incorrect.

Filtration Fraction (FF) is defined by the equation GFR/RPF. Assuming that GFR remains unchanged, a decrease in Hct would lead to a rise in RPF, so filtration fraction would be decreased. Therefore, (C) and (D) are incorrect.
A 70 kg man is administered inulin. Because it is freely filtered and not secreted or reabsorbed, inulin offers an accurate calculation of GFR. Inulin would be found in what amount of this patient’s body water?

a. 8.4 L  

b. 14 L  

c. 16.8 L  

d. 20 L  

e. 40 L

**correct answer:** B

In order to answer this question, you need to know two pieces of information. First of all, you need to remember the 60-40-20 rule related to body weight. 60% of body weight is total body water, 40% of body weight is intracellular fluid and 20% is extracellular fluid. Inulin is found in the extracellular fluid and can be used to measure extracellular fluid volume. To answer this question, it easiest to divide 70 by 10, which tells us that 10% of her body weight is 7 kg. Multiplying this value by 2 would give us 20% of body weight, or 14 kg, which is equivalent to 14 L of water,
You have been working with a couple who has been trying to conceive for 6 months, without success. You schedule the husband for a work-up, which reports that his sperm count is normal but the volume of the ejaculate is low. The concentration of fructose is very low. Which secondary sex gland produces a slightly alkaline, viscous fluid into the semen that is rich in fructose?

a. Ampulla  
b. Bulbourethral gland  
c. Epididymis  
d. Prostate  
e. Seminal vesicles

**Correct answer:** E

The ampulla is the end of the vas deferens.

The bulbourethral glands secrete mucus for lubrication.

The epididymis concentrates and stores sperm for ejaculation.

The prostate gland secretes a fluid rich in citric acid, lipids, zinc, and acid phosphatase.

The seminal vesicles produce fructose, proteins, enzymes, Vitamin C, flavins prostaglandins, phosphorylcholine, and fibrinogen. These comprise about 60% of the volume of semen.
A patient who is expecting her first child comes to your office with test results she would like you to review. Prior to pregnancy, she has been in good health with normal serum laboratory values in the past. Which of the following results would you expect to be elevated in this patient?

a. Blood pressure
b. Creatinine
c. Hematocrit
d. Platelets
e. Thyroid binding globulin

correct answer: E

The hyperestrogenic state of pregnancy causes an increase in thyroid binding globulin (TBG). Placental hormones, including beta HCG, have thyroid-stimulating properties that lead to an elevation in total T3 and T4 and suppression of TSH. Together, these changes lead to a relatively euthyroid state.

During pregnancy, systemic vascular resistance decreases (Choice A), likely due to elevated progesterone levels that cause smooth muscle relaxation. Systolic blood pressure usually decreases by 5 to 10 mm Hg and diastolic blood pressure by 10 to 15 mm Hg. The decrease in blood pressure reaches a nadir at about week 24, at which point it slowly returns to prepregnancy levels at term. The decrease in blood pressure is accompanied by an increase in cardiac output, generally by 30 to 50%.

The glomerular filtration rate (GFR) increases by 50% in early pregnancy and is maintained at this level until delivery. As a result of the increased GFR, blood urea nitrogen and creatinine (Choice B) decrease by approximately 25%.

During pregnancy, plasma volume increases by 50%. Red blood cell levels increase by only 20 to 30%, however, leading to a decrease in hematocrit (Choice C). Similarly, there is a slight decrease in the concentration of platelets (Choice D). This is attributed not only to the increase in plasma volume, but also increase in peripheral destruction. A drop in platelet level below 100 million/mL, however, is not normal and warrants further investigation.
An 18-year-old female presents with abdominal pain determined to be related to ovulation. In reviewing the hormones of the menstrual cycle, you correctly remember that which of the following is responsible for ovulation?

- a. Estrogen
- b. Follicle stimulating hormone (FSH)
- c. Luteinizing hormone (LH)
- d. Progesterone
- e. Prolactin

**Correct answer:** C

LH is released by the anterior pituitary in a spike toward the end of the follicular phase when estrogen levels surge to a critical level. Ovulation occurs as the LH surge causes the follicle to rupture and release the mature ovum. It can be associated with unilateral abdominal pain, also known as mittelschmerz. After ovulation, the granulosa and theca interna cells of the dominant follicle form the corpus luteum under stimulation by LH, marking the beginning of the luteal phase.

FSH (Choice B) is released by the pituitary during the follicular phase and is responsible for stimulating the growth of ovarian follicles.

Estrogen (Choice A) is produced by the developing follicle destined to ovulate. It enhances follicular maturation and increases the production of FSH and LH receptors in an autocrine fashion, thus protecting the dominant follicle from the negative feedback on pituitary FSH secretion caused by the rising estrogen levels.

Progesterone (Choice D) is synthesized by the corpus luteum, in addition to estrogen. It is responsible for preparing the endometrium for implantation of a fertilized ovum. If fertilization occurs, the developing trophoblast synthesizes human chorionic gonadotropin (hCG) to maintain the corpus luteum until the placenta develops. If fertilization fails to occur, the corpus luteum degenerates, progesterone levels fall, and menstruation occurs. The decline in estrogen and progesterone during the luteal phase causes a gradual increase in FSH, thus continuing the cycle.

Prolactin (Choice E) is produced in high levels during pregnancy due to high circulating levels of estrogen. During this time, prolactin fosters the maturation of mammary glands in preparation for lactation. After childbirth, prolactin levels fall unless stimulated by suckling.
A man has a tragic horseback riding accident in which he is paralyzed below T4. Which of the following lung function measures will be decreased?

- a. Tidal Volume (TV)
- b. Inspiratory Capacity (IC)
- c. Inspiratory Reserve Volume (IRV)
- d. Expiratory Reserve Volume (ERV)
- e. Residual Volume (RV)

**Correct answer:** D

TV, IC, and IRV are all volumes that are generated by voluntary contraction of the diaphragm. Since the diaphragm is innervated by C3-C4-C5, inspiratory volumes will be unchanged with injury below C5. Remember TV+IRV=IC. Expiration is normally a passive process, but forceful expiration can be generated by contraction of abdominal muscles. Since the abdominal muscles (rectus abdominis, external and internal obliques) are innervated by T7-T12, injury at T4 will paralyze voluntary abdominal contraction, and eliminate the ability to produce ERV (Answer D). RV is the volume that remains in the lungs after forceful expiration. This volume is dependent on the dead space. In this case the functional dead space would actually increase due to lack of ERV.
A 25-year-old hiker is traveling to the mountains to begin a several week hike. Over the course of several weeks his body will adjust to the high altitude by all of the following EXCEPT:

- Decreasing 2,3 DPG
- Increasing his minute ventilation
- Increasing production of RBCs
- Increasing renal excretion of bicarbonate
- Increasing the number of mitochondria per cell

**Correct answer:** A

At high altitude the body will increase the amount of 2,3 DPG in an effort to unload more bound oxygen from hemoglobin. The relative hypoxia will also cause him to (B) increase his minute ventilation, which will cause a respiratory alkalosis. The kidney will compensate by (D) increasing excretion of bicarbonate. Increased erythropoietin will stimulate production of more RBCs and the body will produce more mitochondria per cell.
A 60-year-old female with a 50-pack-year history of smoking presents complaining of chronic shortness of breath. She brings you her results from a recently-performed pulmonary function test. Which of the following would you most likely expect to see on spirometry?

a. FEV1 decreased, FEV1/FVC decreased, TLC increased, VC decreased
b. FEV1 decreased, FEV1/FVC decreased, TLC increased, VC decreased
c. FEV1 decreased, FEV1/FVC normal, TLC decreased, VC decreased
d. FEV1 increased, FEV1/FVC increased, TLC decreased, VC increased
e. FEV1 increased, FEV1/FVC normal, TLC decreased, VC increased

**correct answer:** A

This set of PFTs is characteristic of obstructive lung disease, such as emphysema. Choice B is characteristic of restrictive lung disease, such as interstitial fibrosis. Choices C, D, and E are not characteristic of either obstructive or restrictive lung disease. Spirometry is performed by having the patient exhale as rapidly and forcibly as possible following a maximum inspiration. The spirometer plots the change in lung volume against time and can help to distinguish obstructive from restrictive lung disease.
A patient is admitted to the hospital with shortness of breath and found to have a pleural effusion on chest X-ray. Thoracentesis is performed and the fluid is sent for laboratory testing. Results of the fluid analysis are: pleural protein 8.0, serum protein 6.5, pleural LDH 500, serum LDH 100. Based on these findings, which of the following is the most likely etiology of the pleural effusion?

a. Bacterial  
b. Congestive heart failure (CHF)  
c. Cirrhosis  
d. Nephrotic syndrome  
e. Pulmonary embolism

**Correct answer**: A

Light's criteria can be used to distinguish exudative and transudative causes of pleural effusions. Based on this criteria, exudative effusions possess at least one of the following characteristics: 1) ratio of pleural fluid protein to serum protein greater than 0.5, 2) ratio of pleural fluid LDH to serum LDH greater than 0.6, and 3) pleural fluid LDH must be greater than 2/3 the upper limit of normal serum LDH. Transudates do not meet any of the criteria. Common causes of exudative effusions include bacterial pneumonia, tuberculosis, and malignancy.

CHF (Choice B), cirrhosis (Choice C), and nephrotic syndrome (Choice D) are all common causes of transudative effusions. Pulmonary embolism (Choice E) can lead to either a transudative or exudative effusion; however, the results of the fluid analysis in this question meet all three of the Light's criteria for an exudative effusion.
A 60-year-old female with a 50-pack-year history of smoking presents complaining of chronic shortness of breath. She brings you the following results of recently-performed pulmonary function tests. What would you most likely expect to see on spirometry?

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b. FEV1 decreased, FEV1/FVC increased, TLC increased, VC increased
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d. FEV1 increased, FEV1/FVC increased, TLC decreased, VC increased
e. FEV1 increased, FEV1/FVC normal, TLC decreased, VC increased

**correct answer: A**

This set of PFTs is characteristic of obstructive lung disease, such as emphysema.

Choice C is characteristic of restrictive lung disease, such as interstitial fibrosis.

Choice B, D, and E are not characteristic of either obstructive or restrictive lung disease.

Spirometry is performed by having the patient exhale as rapidly and forcibly as possible following a maximum inspiration. The spirometer plots the change in lung volume against time and can help to distinguish obstructive from restrictive lung disease.
A normal, healthy, 25-year-old man lives at the beach. His twin brother has been living in a mountain cabin for the past 2 years. Which of the following indices would be expected to be higher in the man living at sea level?

- a. Diameter of pulmonary vessels
- b. Erythropoietin production
- c. Mitochondrial density in a muscle biopsy
- d. Renal bicarbonate (HCO3-) excretion
- e. Respiratory rate

**Correct answer: A**

The correct answer is A. A number of physiologic changes occur in a person living at high altitude. The diminished barometric pressure at high altitude causes alveolar hypoxia and arterial hypoxia. Pulmonary vasoconstriction occurs in response to alveolar hypoxia; therefore, the diameter of the pulmonary vessels would be greater in the brother living at sea level. All the other choices describe physiologic processes that would be enhanced by living at high altitude.

Increased erythropoietin production (choice B), caused by arterial hypoxia, leads to increases in hematocrit in people living at high altitude.

Mitochondrial density increases (choice C) in people chronically exposed to the hypoxemia caused by living at high altitude. At high altitudes, the ventilation rate increases, causing a respiratory alkalosis. The kidney then compensates by increasing the excretion of HCO3- (choice D).

Increasing the rate of respiration (choice E) is a very useful adaptation to the hypoxic conditions of high altitude. The primary stimulus is the hypoxic stimulation of peripheral chemoreceptors.
A researcher is investigating epithelial cell junctions. She injects one cell with a liquid dye and notes that the dye seeps into the adjacent cell. Which epithelial cell junction is most likely to have allowed the dye through to the adjacent cell?

a. Desmosome
b. Gap junction
c. Hemidesmosome
d. Zona adherens
e. Zona occludens

**Correct answer:** B

Gap junctions (choice B) allow adjacent cells to communicate with one another. They are made up of connections with central channels which allow small substances (< 1000 daltons) to pass between cells.

Desmosome (choice A), also known as a macula adherens, is a discrete site of attachment between cells that stabilizes the cell to cell connection; however, it does not allow any substance to pass between the cells.

Hemidesmosomes (choice C) attach the cell to its underlying extracellular matrix and are not involved in cell to cell communication.

Zona adherens (choice D) is made up of actin filaments and E-cadherin and participates in stabilizing the border between cells. It does not allow communication between cells.

Zona occludens (choice E) is a tight junction between cells that prevents substances from diffusing across the intracellular space.

From basal surface: hemidesmosomes (ECM/basement membrane) --> zona adherens --> macula adherens --> zona occludens/tight junction
A 55-year-old woman presents to her primary care physician complaining of increasing difficulty climbing stairs and getting out of chairs over the last several months. She denies fatigue, muscle aches, weight loss, fever, or joint pain. She does not take any medications and denies the use of drugs or alcohol. Physical examination reveals a reddish-purple discoloration and swelling of the eyelid, as pictured. In addition, a scaly, erythematous rash is seen over the knuckles. Which of the following test results would most likely establish the probable diagnosis?

a. Absent anti-nuclear antibodies
b. Decreased erythrocyte sedimentation rate
c. Decreased membrane irritability on electromyography
d. Elevated serum creatine kinase
e. No significant findings on muscle biopsy

Correct answer: D

Dermatomyositis, classified as an idiopathic inflammatory myopathy, presents with proximal muscle weakness that usually begins in the lower limbs. An inability to climb stairs or difficulty rising from a seated position may be the first sign. Dermatomyositis is characterized by skin manifestations, in addition to the myopathy. The violaceous eruption pictured is known as the heliotrope rash, and is often accompanied by eyelid swelling. Other typical findings include Gottron’s sign, an erythematous, scaly eruption found over the metacarpophalangeal and interphalangeal joints, and the shawl sign, a flat, erythematous lesion found over the chest and shoulders in a shawl-like distribution. In addition to clinical history and signs, laboratory values used in the assessment of dermatomyositis include an elevated serum CK (Choice D) and erythrocyte sedimentation rate (Choice B). Electromyography (Choice C) is used to distinguish myopathic and neuropathic causes of muscle weakness and shows increased membrane irritability in dermatomyositis. Muscle biopsy is the definitive test for establishing the diagnosis of inflammatory myopathy and typically reveals myositis.
pt is dark skinned and obese with hyperpigmented skin lesion in axillary area, pt may be predispose to gastric adenocarcinoma
   a. staph areus
   b. acanthosis albicans
   c. nevel
   d. psoriasis
   e. eczema

**Correct answer:** B

Commonly associated with cases with dark skinned obese individuals, you must be wary that they may get GASTRIC adenocarcinoma! You cannot miss this and the NBME won't let you off if you don't know this