F. VISCERAL AND OTHER SYNDROMES OF THE TRUNK APART FROM SPINAL AND RADICULAR PAIN

GROUP XVII: VISCERAL AND OTHER CHEST PAIN

Acute Herpes Zoster (XVII-1)

Code
303.X2d

Postherpetic Neuralgia (XVII-2)

Code
303.X2e

The syndromes of herpes zoster and postherpetic neuralgia are similar in all regions and are normally unilateral and limited to one or two dermatomal segments. For a general description of them see 11-4 and 11-5.

Postinfectious and Segmental Peripheral Neuralgia (XVII-3)

Definition
Paroxysmal pain in the distribution of an intercostal nerve commonly associated with cutaneous tenderness in the affected dermatome.

Site
In the distribution of spinal nerve roots or trunks (if segmental neuralgia); in the distribution of the intercostal nerves; or in the distribution of the posterior primary division of the nerve trunk (if peripheral neuralgia).

System
Peripheral nervous system.

Main Features
Pain Quality: sharp or burning pain, usually intermittent, often precipitated by lateral movements of trunk or vertebral column. Associated with tenderness at points of exit of the nerve from a deep to a more superficial plane, e.g., at mid-axillary or parasternal lines. Post-traumatic intercostal neuralgia often has continuous pain with exacerbation.

Etiology
Neuralgic pains may be due to postinfectious radiculitis, osteoarthritic spurs, other spinal lesions, trauma, toxic and metabolic lesions, etc. In acute cases they are most often precipitated by upper respiratory infection or trauma. In chronic cases bad body mechanics, lordosis or scoliosis, trauma, and arthritis are the most common causes. Primary neuralgia occurs rarely.

Differential Diagnosis

From neuralgias attributable to specific causes, or described above.

Code

306.X2	Postinfectious
306.X8	Unknown
306.X1 or 303.Xla	Post-traumatic

Angina Pectoris (XVII-4)

Definition
Pain, usually constricting, and a heavy feeling in the chest, related to ischemia of the myocardium without myocardial necrosis.

Site
Pain classically is in the precordium, although radiation to the arms and hands is common, particularly to the medial aspect of the left arm. Pain may also radiate up into the sides of the neck or jaw or into the back or epigastrium.

System
Cardiovascular system.

Main Features
Prevalence: common in middle and older age groups, males more than females. *Pain Quality:* the pain tends to be dull, crushing, or constricting and heavy. It is frequently precipitated by stress, either physical or psychological. It usually lasts a few minutes but can be prolonged or intermittent, lasting hours or occasionally longer.

Associated Symptoms
As noted, pain is aggravated by stress and relieved promptly by rest or nitroglycerin. Frequently patients also experience breathlessness, sweating, nausea, and belching.

Signs and Laboratory Findings
Frequently there are no objective findings but patients may at the time demonstrate a tachycardia, a mitral regurgitant murmur of papillary muscle dysfunction, an S3 or S4, and reversed splitting of the second heart sound. Laboratory testing may show characteristic electrocardiographic changes, most commonly ST segment depression. Between pains, a stress electrocardiogram may show ST depression with exercise. Coronary angiography may show typical atherosclerotic narrowing of the coronary arteries.

Usual Course
Anginal pain typically is brief and intermittent, brought on by exertion or stress and relieved by rest and nitroglycerin. It may remain stable over many years, or may become “atypical” or accelerate to “preinfarction (or “unstable”) angina.”

Complications
Arrhythmia and myocardial infarction may occur.

Social and Physical Disability
If angina is brought on by little extra stress, there is serious reduction in the work capacity. If the patient is particularly fearful, angina can cause interruption of normal psychological function as well. The big concern is usually fear of progression to sudden death or myocardial infarction, though limitation of

activity level may also be a serious threat.

Pathology
A list of risk factors predisposing individuals to atherosclerotic heart disease continues to develop but includes age, sex, hypertension, smoking, family history, hyperlipidemia, obesity, sedentary life-style, diabetes, etc. Superimposed on atherosclerotic coronary artery narrowing, such factors as increased cardiac oxygen demand, decreased flow related to coronary artery spasm, or arrhythmias may be contributory.

Summary of Essential Features and Diagnostic Criteria
Crushing retrosternal chest pain brought on by stress (physical or psychological) and relieved by rest and nitroglycerin, with ST depression on ECG but no evidence of infarction on sequential ECGs and cardiac enzymes.

Code
324.X6
224.X6 If mostly in the arms

Myocardial Infarction (XVII-5)

Definition
Pain, usually crushing, from myocardial necrosis secondary to ischemia.

Site
Retrosternal area with radiation to arms, neck, jaw, epigastrium.

System
Cardiovascular system.

Main Features
Prevalence: common in middle and older age groups, especially males. *Pain Quality:* the pain is dull, central, retrosternal, and heavy or crushing. Usually it is very severe and lasts several hours or until relieved by morphine. It may diminish slowly over a day or two.

Associated Symptoms
Breathlessness, sweating, nausea and vomiting, apprehension, and lightheadedness are common.

Signs and Laboratory Findings
Physical examination may be normal but may show hypertension, S3 or S4 gallop rhythm, and papillary muscle dysfunction with a mitral regurgitant murmur, as well as signs of forward or backward cardiac failure.

Laboratory abnormalities include elevation of cardiac enzymes such as CPK, LDH, and SGOT, classical sequential ECG changes (ST elevation and the development of Q waves), and abnormal radionuclide heart scan.

Usual Course
In patients surviving myocardial infarction the severe pain tends to diminish and disappear over several hours to a day or two. Often the patient is then pain free, although recurrent pain may represent angina or reinfarction.

Complications
Sudden cardiac death, arrhythmias, congestive heart failure, cardiogenic shock, post-myocardial infarction syndrome, pericarditis, septa] perforation, valve cusp rupture, mural thrombus and embolism, myocardial aneurysm, deep vein thrombosis, and pulmonary embolism.

Social and Physical Disability
Myocardial infarction is a major cause of death and disability. Recovery frequently takes several months, and physical and psychological complications may prolong recovery and affect not only the patient but family members, friends, and employers. The significance of the heart as the source of life makes interpretation of this type of pain particularly threatening.

Pathology
The main pathogenic process is atherosclerosis of the coronary arteries. Other factors such as coronary artery spasm or arrhythmias, or decreased blood volume, or decreased total peripheral resistance may also be significant as "last straws." The risk factors mentioned with Angina—sex, age, hypertension, smoking, Type A personality, etc.—are important predisposing factors.

Differential Diagnosis
Angina pectoris, dissecting aneurysm, pulmonary embolism, esophageal spasm, hiatus hernia, and pericarditis.

Summary of Essential Features and Diagnostic Criteria
Crushing retrosternal chest pain with myocardial necrosis as evidenced by ECG and enzyme changes.

Code
321.X6
221.X6 If in the anus

Pericarditis (XVII-6)

Definition
Pain, often sharp, arising from inflammation of the pericardium.

Site
The pain is classically in the precordium but may radiate through to the midthorax posteriorly or follow the pattern of angina, or to the superior border of the trapezius muscles.

System
Cardiovascular system.

Main Features
Most cases are acute, and this is particularly true of pericarditis causing pain. Pain may be severe and pleuritic. It may be aggravated by swallowing. It may be steady, crushing substernal pain. It may occasionally be synchronous with the heartbeat.

Associated Symptoms
Weight loss, fatigue, and fever are common especially in chronic cases.

Signs and Laboratory Findings

Leaning forward may reduce the pain. The hallmark sign is a triphasic pericardial friction rub. If there is a significant effusion, heart sounds may be decreased and a paradoxical pulse may be elicited.

Laboratory signs include a "water bottle" configuration on chest X-ray if there is an effusion, as well as changes in fluoroscopy, echocardiography, or angiography. Even without an effusion the electrocardiogram may show typical changes, ST elevation, or inverted T waves.

Usual Course
The course varies depending on the etiology and may range from being acute to chronic.

Complications
May interfere with cardiac output.

Social and Physical Disabilities
Probably only significant in chronic cases where weight loss and generalized debility are part of the syndrome.

Etiology
A wide range of etiologies can cause pericarditis and its subsequent pain. The most treatable causes are infections, collagen, vascular, and drug-induced effects. Postmyocardial injury is also an important etiology.

Summary of Essential Features and Diagnostic Criteria
Sharp retrosternal pain aggravated by breathing and relieved by leaning forward, with auscultation revealing a friction rub, ECG showing ST elevation or T wave inversion, and echocardiogram showing an echo-free pericardial space.

Differential Diagnosis
Angina, myocardial infarction, pulmonary embolism, hiatus hernia, and esophageal spasm, etc.

Code

323.X2	Known infection
323.X3	Unknown infective cause
323.X1	Trauma
323.X4	Neoplasm
323.X5	Toxic

Aneurysm of the Aorta (XVII-7)

Definition
Pain from an abnormal widening of the aorta.

Site
Central chest pain which may radiate to the mid-scapular region.

System
Vascular system.

Main Features
Deep, diffuse, aching central chest pain is associated with large aneurysms. If dissection occurs, sudden

and severe pain occurs, maximal at onset.

Associated Symptoms
Acute congestive heart failure may develop.

Signs and Laboratory Findings
A discrepancy may develop between pulses or blood pressures in the two arms. A new aortic regurgitant murmur may develop. A neurological impairment may develop. Chest X-ray may show widening of the superior mediastinum. Aortography may demonstrate a false lumen.

Usual Course
If there is a large aortic aneurysm, there can be chronic dull, central chest aching. If dissection occurs, an acute medical and surgical emergency has developed.

Complications
Chronic complications include pain and emboli. Acute complications include acute aortic valvular incompetence, occlusion of major vessels, hypotension, and death.

Social and Physical Disability
The main problems with aortic aneurysms are life and death considerations.

Pathology
"True" aneurysms involve all three layers-intima, media, and adventitia. "False" aneurysms involve disruption of the inner and medial segments so that the wall of the aneurysm consists only of adventitia and/or perivascular clot.

In the past, syphilis was the main cause. Cystic medial necrosis is a major cause of dissection. Arteriosclerosis is a major cause. Occasionally Marfan's syndrome is the cause. Hypertension is important and so is trauma.

Summary of Essential Features and Diagnostic Criteria
A rare cause of chronic chest pain with a wide superior mediastinum on chest X-ray. A dramatic cause of excruciating acute pain with importance because of medical and surgical therapies available.

Differential Diagnosis
Angina, pulmonary diseases, and thoracic disk disease.

Code
322.X6 Chronic aneurysm

Chronic aneurysm
If the pain assumes a thoracic spinal pattern (although of visceral origin), code according to X-7.2 as 322.X6a.

Diseases of the Diaphragm (XVII-8)

Definition
Pain from the diaphragm related to irritation of the diaphragmatic nerves by a disease process above the diaphragm, in the diaphragm (rare), or below the diaphragm.

Site
Diaphragmatic pain is deep and difficult to localize. Noxious stimulation may affect phrenic nerve sensory fibers C3, C4, and C5 and therefore is often felt at the shoulder tips and along the upper border of the trapezius muscle, or it may affect the intercostal nerves T6, T7, T8, and T9 with radiation of pain into the anterior chest, the upper abdomen, and the corresponding region of the back.

System
The system is musculoskeletal, cardiac, pulmonary, or intestinal depending upon the disease.

Main Features
The pain is deep, dull, poorly localized, and non-specific if it involves only the central chest and upper abdomen and upper back, but becomes better identified if there is shoulder tip radiation as well.

Associated Symptoms
These symptoms depend most on the underlying pathology, so that if there is pulmonary pathology, respiratory symptoms may be prominent. Likewise, if the basic disease is gastrointestinal or subphrenic, gastrointestinal complaints are most likely associated. Hiccoughs may be present.

Signs and Laboratory Findings
Frequently there are no physical findings, but if there are, the most classic would be elevation of a hemidiaphragm. Laboratory testing might show elevation of the diaphragm on chest X-ray, abnormal movement of the diaphragm on fluoroscopy, a pleural effusion on chest X-ray, an echo-free space on abdominal ultrasound, a space between liver and heart on radio-nuclide imaging, or a space beneath the diaphragm on CT scan or nuclear magnetic resonance scanning.

Usual Course
There is usually a specific therapy once the etiology is determined, but a considerable time may elapse before a conclusive diagnosis is reached.

Complications
Depend on the underlying cause.

Social and Physical Disability
These relate partly to the underlying disease process and partly to the vagueness of understanding of the cause of pain. This latter can be extremely frustrating to doctor and patient alike.

Etiology
Although a wide range of causes can cause disease affecting the diaphragm, the most important are infections and neoplasms.

Summary of Essential Features and Diagnostic Criteria
Abdominal pain in epigastrium with radiation to central chest, posterior midthorax and shoulder tip(s), with evidence of space-occupying lesions above or below the diaphragm.

Differential Diagnosis
Involves a wide range of cardiac, pulmonary, musculoskeletal, and gastrointestinal uses.

Code

423.X2	Infection: chest or pulmonary source
423.X4	Neoplasm: chest or pulmonary source
433.X2	Musculoskeletal

453.X2	Infection: gastrointestinal source
453.X4a	Neoplasm: gastrointestinal source
453.X6	Cholelithiasis

Carcinoma of Esophagus (XVII-9)

Definition
Pain due to malignant disease of the esophagus resulting from malignant transformation of either the squamous epithelium of the upper esophagus or the mucosa of the lower esophagus.

Site
Retrosternal pain, extending sometimes to the back.

System
Gastrointestinal system.

Main Features
This is a relatively uncommon tumor in the Western World but has localized areas of high incidence, especially in Iraq and Iran among the Kurds. Pain is not usually a prominent feature. The presenting symptom is usually dysphagia without pain, which usually occurs only when the cancer extends beyond the esophagus. At that point dysphagia and retrosternal pain may become continuous and radiate through the back.

Associated Symptoms
Dysphagia is the major symptom; others include regurgitation and recurrent pneumonia.

Signs and Laboratory Findings
Evidence of weight loss and cervical lymphadenopathy, particularly deep to the sternomastoid. Chest X-ray may show a dilated esophagus; barium swallow, a narrowing of the esophageal lumen; iron-deficiency anemia.

Usual Course
Unless the tumor is removed, the patient will become obstructed.

Complications
Esophageal obstruction, erosion into a bronchus, bronchoesophageal stricture, erosion into aorta with catastrophic hemorrhage.

Social and Physical Disability
If the tumor is inoperable and the patient cannot eat, a plastic tube can be passed through the tumor or a feeding jejunostomy performed.

Pathology
Chronic ingestion of carcinogens in certain areas of the world, e.g., Kurdistan and Lake Victoria, East Africa. Smoking-chronic disorders of esophagus, e.g., achalasia, Barrett's esophagus.

Summary of Essential Features and Diagnostic Criteria
Presents with dysphagia with pain as a late feature. Diagnosed by barium swallow and esophagoscopy with biopsy or cytology.

Differential Diagnosis
Benign stricture, achalasia.

Code
353.X4

Slipping Rib Syndrome (XVII-10)
(also known as Clicking Rib Syndrome and Rib Tip Syndrome)

Definition
Chronic pain at the costal margin which may mimic visceral pain.

Site
Eighth, ninth, or tenth rib cartilages, one or more rib cartilages being involved. The condition may be bilateral.

Main Features
Prevalence: fairly common. *Age of Onset:* 15-60 years. *Sex Ratio:* more common in females (3F:1M). *Quality: a* constant dull ache or a sharp stabbing pain which may itself be followed by a dull ache. *Time Pattern:* the pain may last from several hours to many weeks, and some patients have constant pain.

Aggravating Factors
Movement, especially lateral flexion and rotation of the trunk. Rising from a sitting position in an armchair is often a particularly painful stimulus.

Signs
Manipulation of the affected rib and its costal cartilage will exactly reproduce the presenting pain.

Usual Course
Some cases may resolve spontaneously, but most patients have symptoms permanently.

Relief
Restriction of movement may give relief.

Complications
Depression and anxiety. Patients may be misdiagnosed and undergo unnecessary investigations and even inappropriate surgical procedures.

Social and Physical Disability
Physical activities are often restricted by pain or fear of provoking an exacerbation.

Pathology
No specific histological changes identified. Cause of pain is presumed to be irritation of intercostal nerve by adjacent hypermobile rib cartilage.

Summary of Essential Features and Diagnostic Criteria
A fairly common condition which should be considered in any patient complaining of upper abdominal pain. The diagnosis is clinical and should be made only when the patient's symptoms are exactly reproduced by manipulation of the appropriate rib or ribs. An intercostal nerve block with local anesthetic

may produce confirmatory evidence where the clinical findings are equivocal.

Treatment
Reassure patient-this may be sufficient for some patients who do not have severe pain. If severe pain persists, then the offending costal cartilage should be excised.

Differential Diagnosis
Biliary tract pathology, duodenal and gastric ulceration.

Code
333.X6

References
Copeland GP, Machin DG, Shennan JM. Surgical treatment of the slipping rib syndrome. Br J Surg 1984;71:522–3.

Holmes JF. Slipping rib cartilage. Am J Surg 1941;54:326–38.

Postmastectomy Pain Syndrome: Chronic Nonmalignant (XVII-11)

Definition
Chronic pain commencing immediately or soon after mastectomy or removal of a lump, affecting the anterior thorax, axilla, and/or medial upper arm.

Site
Anterior thorax, axilla, medial upper arm; usually one side only.

System
Peripheral nervous system.

Main Features
Prevalence: infrequent. *Age of Onset:* any age. *Sex:* females. *Pain Quality:* often burning, intensified by touch or clothing. *Time Pattern:* constant; unremitting to analgesic. *Intensity:* moderate to severe. *Duration:* years.

Associated Symptoms
The patient may be unable to tolerate a prosthesis, clothing, or touch.

Signs
Increased response to touch; hyperesthesia and allodynia to skin stroking or skin traction. Reduction in appreciation of pinprick, cold, and touch related to the incision and upper arm.

Usual Course
May remain intractable to physical measures. Commonly responds to amitriptyline. May also respond to ointments based on capsaicin.

Complications
Can be compounded by emotional stress, recurrence of disease.

Social and Physical Disability

Impairment of social, occupational, and sexual activities.

Pathology
None known.

Summary of Essential Findings and Diagnostic Criteria
Pain commencing postoperatively, usually immediately, at the site of the mastectomy, without objective evidence of local abnormality.

Allodynia over widespread areas of the chest or arm, or both; sensory loss over anterior chest or arm, or both.

Differential Diagnosis
Herpes zoster, local infection, radiation necrosis in ribs, recurrent neoplasm.

Code
303.X9

Late Postmastectomy Pain or Regional Carcinoma (XVII-12)

Definition
Shooting, jabbing, or burning pain commencing more than three years after the initial treatment for cancer of the breast and due to local metastases.

Site
Spine, thorax at site of cancer, arms.

Main Features
Prevalence: fairly common. *Age: 35* and upward. *Sex Ratio:* most common in females. *Pain Quality:* varies according to etiology. With skeletal secondary deposits, pain is increased with movement. Shooting or jabbing pain occurs with brachial plexus lesion, usually spontaneously, sometimes with paresthesias. Burning, shooting, and numb feelings are found with brachial plexus damage from radiation. *Intensity:* moderate to severe. *Duration:* less than 12 months, due to short life expectancy.

Associated Symptoms
Weakness and reduced range of movement of the ipsilateral limb.

Signs
Ipsilateral Homer's syndrome; lower brachial plexus signs, e.g., anesthesia, muscle weakness, and wasting; chest wall or axillary or supraclavicular recurrent disease. Bony tenderness to percussion.

Usual Course
With skeletal secondaries and brachial plexus damage, the course is usually progressive deterioration. However, with radiation damage to the brachial plexus, the course is more protracted, with onset more than five years after treatment and long survival.

Complications
Patients with skeletal, visceral, and brachial plexus damage have a short survival of less than one year. Radiation damage is a progressive disorder with disability and long survival.

Social and Physical Disability
Moderate impairment of social and occupational activity, with depression related to chronic illness.

Pathology
Local skin, subcutaneous, skeletal, or visceral metastatic disease; with recurrent disease there is local lymphatic spread, and extradural and brachial plexus involvement. Radiation damage to the brachial plexus is more common in patients who have received repeated or excessive doses of radiation, and in such patients, telangiectasia may be present in the skin with pigmentation and signs of radiation arthritis.

Diagnostic Criteria
Pain arising more than three years after mastectomy for cancer, at the above sites. Objective evidence of recurrent disease.

Differential Diagnosis
Herpes zoster; pleurisy related to infection; and second tumor, e.g., Pancoast's tumor with brachial plexus involvement.

Code
307.X4

Post-thoracotomy Pain Syndrome (XVII-13)

Definition
Pain that recurs or persists along a thoracotomy scar at least two months following the surgical procedure.

Site
Chest wall.

Systems
Skeletal and nervous systems.

Main Features
Pain following thoracotomy is characterized by an aching sensation in the distribution of the incision. It usually resolves in the two months following the surgery. Pain that persists beyond this time or recurs may have a burning dysesthetic component. There may also be a pleuritic component to the pain. Movements of the ipsilateral shoulder make the pain worse.

Associated Symptoms
If the thoracotomy was done for tumor resection and there was evidence of pleural or chest wall involvement at the time of surgery, it is likely that the pain is due to tumor recurrence in the thoracotomy scar.

Signs and Laboratory Findings
There is usually tenderness, sensory loss, and absence of sweating along the thoracotomy scar. Auscultation of the chest may reveal decreased breath sounds due to underlying lung consolidation or a malignant pleural effusion. A specific trigger point with dramatic pain relief following local anesthetic injection suggests that the pain is benign in nature and due to the formation of a traumatic neuroma. A CT scan through the chest is the diagnostic procedure of choice to establish the presence or absence of recurrent tumor.

Usual Course

If the pain is due to traumatic neuromata, it usually declines in months to years and can be relieved by antidepressant-type medications and anticonvulsants. If the pain is due to tumor recurrence, some relief may be obtained by an intercostal nerve block or radiation therapy.

Complications
Immobility of the upper extremity because of exacerbation of the pain may result in a frozen shoulder. Aggressive physiotherapy is necessary to prevent this complication.

Pathology
For benign disease, the pathology is that of neuroma formation. If there is an underlying malignancy, there is tumor infiltration of the intercostal neurovascular bundle.

Summary of Essential Features and Diagnostic Criteria
Persistent or recurrent pain in the distribution of the thoracotomy scar in patients with lung cancer is commonly associated with tumor recurrence. CT scan of the chest is the diagnostic procedure of choice to demonstrate this recurrence.

Differential Diagnosis
Epidural disease and tumor in the perivertebral region can also produce intercostal pain if there is recurrent disease following thoracotomy.

Code

303.Xld	Neuroma
333.X4a	Metastasis

Internal Mammary Artery Syndrome (XVII-14)

Definition
Burning anterior chest pain commencing immediately or soon after coronary artery bypass surgery in which the internal mammary artery has been used for grafting.

Site
Anterior thorax, usually left side and occasionally bilaterally (always at the site of the graft).

System
Peripheral nervous system.

Main Features
Burning pain across a well-circumscribed area defined by the sternum medially, the intercostal junction at T2 or T3 superiorly, the intercostal junction at T5 or T6 inferiorly, and approximately the nipple line laterally. It is most frequently associated with sharp, spontaneous pains radiating to the chest, axilla, or neck. The pain may be mild, moderate, or intense.

Associated Symptoms
The patients usually do not tolerate contact with clothing or the water of the shower. Occasionally the pain is confused with angina.

Signs and Laboratory Findings
While the area is anesthetic or hypoesthetic, most patients present with troublesome allodynia and also severe tenderness on palpation of the sternum and the costosternal junctions at the site of the harvesting of the graft. Most patients will continue to demonstrate slow healing at the site of the median sternotomy.

An active bone scan may be found up to 4 years after surgery due to compromise of the sternal blood supply as a result of harvesting the internal mammary artery.

Usual Course
Without treatment the pain may decrease in intensity during the first year post surgery, may remain the same, or may become intractable. Thoracic sympathetic ganglia blocks may significantly reduce pain, allodynia, and bone tenderness but only temporarily.

Complications
Pain can be compounded by emotional stress and suspicion of recurrence of heart disease.

Social and Physical Disability
Depending on the degree of discomfort, impairment ranges from negligible to serious.

Pathology
Trauma or resection of the anterior intercostal nerve.

Diagnostic Criteria
Burning pain, numbness, hyperesthesia and deep bone tenderness are almost all simultaneously present at the area of harvesting of the graft.

Relief
The application of TENS/desensitization may be very beneficial within the first few months after surgery. A combination of TENS and tricyclic medication should be tried subject to consideration of the effects of tricyclic antidepressants on the underlying heart disease. Patients may benefit from reassurance that this pain does not arise from recurrent heart disease.

Differential Diagnosis
Ischemic heart pain, costochondritis, hyperesthesia from the scar.

Code
303.X1f

Reference
Mailis A, Chan J, Basinski A, Feindel C, Vanderlinden G, Taylor A, Flock D, Evans D. Chest wall pain after aortocoronary bypass surgery using internal mammary artery graft: a new pain syndrome? Heart Lung 1989;18:553–8.

Tietze's Syndrome— Costo-Chondritis (XVII-15)

Code
332.X6

Fractured Ribs or Sternum (XVII-16)

Code
335.X1

Xiphoidalgia Syndrome (XVII-17)

Code
494.X8

Reference
Bonica JJ. The Management of Pain, Vol. 2. Philadelphia: Lea & Febiger; 1990, p. 1129–30.

Carcinoma of the Lung or Pleura (XVII-18)

Code
323.X4a

GROUP XVIII: CHEST PAIN OF PSYCHOLOGICAL ORIGIN

Muscle Tension Pain (XVIII-1)

Definition
Virtually continuous pain in the thorax, due to sustained muscle contraction and related to emotional causes.

Site
Either symmetrical, more often in the posterior thoracic region, or precordial.

System
Central nervous system (psychological and social).

Main Features
Tension pain is rare in the posterior thoracic region compared with tension headache (perhaps one-tenth or less of the frequency of the latter). Precordial pain is more common, often associated with tachycardia or a fear of heart disease. The exact prevalence is unknown. The other features of these pains are the same as for muscle tension pain in general (I11-1, 2). See also 1-16.

Code
333.X7h

Delusional Pain (XVIII-2)

See the general description of delusional pain (1-16). Perhaps more frequent in the precordial region.

Code
31X.X9d

Conversion Pain (XVIII-3)

See the description of conversion pain in general (1-16). Most frequent in precordium; may be associated with tachycardia and fear or conviction of heart disease being present. Often mimics angina, but without adequate evidence of organic disease.

Differential Diagnosis
As for conversion pain in general and angina pectoris.

Code
31 X.X9e

With Depression (XVIII-4)

See 1-16 for general description.

Code

31X.X9f

GROUP XIX: CHEST PAIN REFERRED FROM ABDOMEN OR GASTROINTESTINAL TRACT

Subphrenic Abscess (XIX-1)

Definition
Pain, often referred to the shoulder, from a collection of pus under the diaphragm.

Site
Upper abdominal pain and tenderness along the costal margins. Shoulder pain may be present.

System
Gastrointestinal.

Main Features
Deep, dull and often poorly localized pain in epigastrium with tenderness beneath the rib margin. Shoulder tip pain often occurs also. Often follows intra-abdominal surgery, especially with perforated viscus. May follow closed blunt trauma.

Associated Symptoms
Fever, malaise, weight loss, hiccoughs.

Signs and Laboratory Findings
These may be vague. The patient may be febrile and cachectic. There may be a pleural effusion or lack of diaphragmatic movement. There may be tenderness to percussion or to palpation of the upper abdomen. White blood cell count and erythrocyte sedimentation rate may be elevated. Chest X-ray may show pleural effusion or elevated hemi-diaphragm. Abdominal ultrasound or CT scan may reveal the collection of pus.

Usual Course
Treatment with antibiotics with or without surgery usually leads to resolution. There may be a prolonged phase of prediagnosis.

Complications
Prolonged fever and weight loss. May lead to death. Chronic pain. Septic shock.

Social and Physical Disability
May lead to usual effects both of chronic sepsis and chronic pain.

Etiology
Most common organisms are E. *colt,* non-group A streptococci, staphylococci, Klebsiella-enterobacter, and anaerobes.

Differential Diagnosis
A wide range of upper gastrointestinal diseases.

Summary of Essential Features and Diagnostic Criteria
Chronic illness often after abdominal surgery with fever and abdominal pain, often with shoulder tip radiation.

Code

353.X2 Thorax
453.X2a Abdomen

Herniated Abdominal Organs (XIX-2)

Definition
Pain related to the protrusion of an abdominal organ through the normal containing walls of the abdomen.

Site
Pain can be related either to the organ herniating or the walls of the orifice. For hiatus hernias the main pain is epigastric.

System
Gastrointestinal system.

Main Features
Burning epigastric pain (or retrosternal pain, or both), often following eating or lying recumbent.

Associated Symptoms
The patient may also complain of chest pain similar to angina, right upper quadrant abdominal pain similar to that in cholelithiasis, epigastric pain like that in peptic ulcer disease, abdominal bloating and air swallowing.

Signs and Laboratory Findings
There are usually no physical findings. Radiographic techniques will show evidence of abdominal viscera in places they are not supposed to be, such as gastric mucosa above the diaphragm or colon above the diaphragm.

Usual Course
Pain typically is intermittent and aggravated by certain foods, aspirin, alcohol, bending over or straining, abdominal pressure or tight clothing, and carbonated beverages. Likewise, more esophageal reflux may occur with caffeine or nicotine, which relax the lower esophageal sphincter.

Complications
Pain and gastrointestinal upset.

Social and Physical Disability
May lead to chronic complaint, usually not too severe.

Etiology
Traumatic and congenital or degenerative weaknesses in the diaphragm are of key etiologic significance, although the exact cause is often obscure.

Summary of Essential Features and Diagnostic Criteria
Epigastric discomfort and esophageal reflux are key symptoms, with radiographic or endoscopic evidence of extra-abdominal organs.

Differential Diagnosis
Angina, cholelithiasis, acid-pepsin disease without hernias, and pancreatitis, etc.

Code

355.X6	Thoracic pain
455.X6	Abdominal pain

Esophageal Motility Disorders (XIX-3)

Definition
Attacks of severe pain, usually retrosternal and midline, due to a diffuse disorder of the esophageal musculature with severe attacks of spasm and/or failure of relaxation of the cardiac sphincter.

Site
Pain is usually well localized to the midline behind the sternum, between the epigastrium and the suprasternal notch.

System
Gastrointestinal system.

Main Features
Prevalence: uncommon. *Sex Ratio:* males and females equally affected. *Age of Onset:* occurs in young adults and middle aged. *Pain Quality:* sudden onset of pain, usually sharp and stabbing, spasmodic and severe, at times excruciating, lasting from 30 seconds to a few minutes, and leaving a residue of retrosternal soreness. The bouts are usually infrequent. Air swallowing and belching are common, and the pain is aggravated by swallowing.

Associated Symptoms
Dysphagia occurs in patients with achalasia of the lower esophageal sphincter. There is a sensation of the food sticking in the lower part of the esophagus. With the aid of gravity, the weight of the food causes the sphincter to open when the patient rises from the chair, and the sticking sensation disappears. Relieving factors include smooth muscle dilatation agents such as glyceryl trinitrate or amyl nitrite, which may relieve the pain.

Signs and Laboratory Findings
Patients usually point out their pain with one finger. Gastroscopy, barium swallow, cine-esophagoscopy or esophageal manometry may show evidence of increased or asynchronous esophageal motility. A barium swallow may show disordered esophageal contractions with or without `spasm' or esophageal dilatation. The cardiac sphincter may remain closed until a large amount of barium fills the esophagus, when it will suddenly open. In patients with prolonged achalasia the esophagus may contain foreign material, which is undigested food. Esophageal manometry will show disordered motility with a lack of normal peristalsis and occasional high-pressure contractions or `spasm.' In patients with achalasia the cardiac sphincter will fail to relax normally following swallowing, although sphincter pressure is normal. Special pressure devices in the esophagus for 24 to 48 hours may pick up very high pressure contractions, which may be related to the pain.

Usual Course
Pain tends to be severe and episodic. It may vary from very occasional to cyclic or be continuous throughout the day. Anxiety and eating may aggravate it. Most patients with motility disorders run a benign course with occasional attacks of pain. Occasionally the symptoms progress to the point where the patient has to undergo active therapy. In contrast, patients with achalasia usually progress to the point where they require definitive treatment.

Complications
If the pain is severe, it may lead to anorexia and weight loss. Vomiting may be a problem. Patients with achalasia can develop aspiration pneumonia from retained esophageal contents. The incidence of esophageal cancer in those patients is slightly increased.

Social and Physical Disability
Severe pain may restrict normal activities and be socially disabling.

Pathology
This is mainly a physiologic rather than a pathologic problem. Stress may be an important contributing factor. There is frequently a positive family history.

Summary of Essential Features and Diagnostic Criteria
This syndrome consists of short attacks of acute severe retrosternal pain which may be relieved by nitrites, with or without dysphagia. The diagnosis is made with a combination of barium swallow appearances and disordered esophageal motility and normal mucosal appearances on esophagoscopy.

Differential Diagnosis
Pericarditis, pulmonary embolism, angina pectoris, dissecting aneurysm, tertiary esophageal contractions in the elderly, and carcinoma of the esophagus.

Code
356.X7

Esophagitis (XIX-4)

Definition
Pain due to inflammation of the esophageal mucosa.

Site
Retrosternal or epigastric pain, depending on the etiology, e.g., upper esophagus, monilial; lower esophagus, acid reflux.

Main Features
Prevalence: common, especially in middle aged and obese. *Sex Ratio:* more common in women. *Pain Quality:* burning retrosternal pain, especially at night if lying flat, or on bending over. *Time Pattern:* may last minutes or hours.

Associated Symptoms
Aggravated by very hot or cold drinks, acidic drinks, alcohol, or strong coffee. Relieved by antacids or food.

Signs and Laboratory Findings
No physical findings. There may be iron-deficiency anemia and positive occult blood tests.

Usual Course
Chronic, intermittent, rarely constant.

Complications
Chronic occult GI bleeding, stricture of lower esophagus.

Social and Physical Disability
Unable to tolerate certain foods, unable to sleep flat in bed.

Pathology
Peptic: Dysfunction of cardiac sphincter results in intermittent regurgitation of gastric acid contents into lower esophagus when intragastric or intra-abdominal pressure is increased and aided by gravity.
Monilial: Commonly secondary to immunosuppression and corticosteroids.

Summary of Essential Features and Diagnostic Criteria
Burning retrosternal pain from esophageal inflammation.

Code

355.X2	Monilial
355.X3a	Peptic

Reflux Esophagitis with Peptic Ulceration (XIX-5)

Definition
Retrosternal burning chest pain due to acid reflux causing inflammation and ulceration.

Site
Typically retrosternal midline pain radiating from behind the xiphisternum up as far as the neck.

System
Gastrointestinal system (esophageal mucosa).

Main Features
Prevalence: common in young adults and middle age group, starting in third decade. *Sex Ratio:* more common in females, especially in the obese or during pregnancy. *Time Pattern:* bouts of pain occur often after postural changes such as bending over or lying down. They also may be associated with a sour taste or waterbrash. *Intensity:* attacks are usually mild, except with ulceration, where they are very severe and last minutes to hours. With ulceration, pain may be continuous.

Associated Symptoms
Sour taste, waterbrash.

Aggravating Factors
Certain postures such as bending over, sitting in a slumped position, or lying down; very hot or cold drinks; acidic drinks. Relieved by antacids.

Signs and Laboratory Findings
The only abnormal findings are appearances of esophagitis (reddening or hemorrhagia mucosa) or of actual ulceration on esophagoscopy. Esophageal motility studies may show a decrease in cardiac sphincter pressure, a pH probe may detect acid reflux, and the pain may be reproduced by the infusion of 0.1 N hydrochloric acid proximal to the cardiac sphincter.

Usual Course
In the majority of patients the symptoms persist intermittently for years. In pregnant women they usually disappear after childbirth, except in the obese patients.

Pathology
Changes in the lower esophageal mucosa may vary from the mildest changes with blunting of the rete papillae to severe hemorrhage inflammation with ulceration and loss of mucosa.

Complications
Patients with ulceration may develop a stricture in the region of the ulcer which can cause dysphagia. Rarely a malignancy may develop in the area of chronic esophagitis.

Summary of Essential Features and Diagnostic Criteria
Esophagitis with nonmalignant ulceration presents with retrosternal pain especially on bending or lying down, or on drinking very hot or cold fluids or eating acidic foods. The diagnosis is made on the history, esophagoscopy, and esophageal motility studies.

Differential Diagnosis
Monilial esophagitis, herpetic esophagitis, foreign body in wall of esophagus, Crohn's disease.

Code
355.X3b

Gastric Ulcer with Chest Pain (XIX-6)

Code
355.X3c

Duodenal Ulcer with Chest Pain (XIX-7)

Code
355.X3d

Thoracic Visceral Disease with Pain Referred to Abdomen (XIX-8)

See Pericarditis (XVII-4) and Diaphragmatic Hernia (XIX-2).

Specific Disease-Associated Abdominal and Pelvic Pain

GROUP XX: ABDOMINAL WALL PAIN

Acute Herpes Zoster (XX-1)

Code
403.X2d

Postherpetic Neuralgia (XX-2)

Code
403.X2b

Segmental or Intercostal Neuralgia (XX-3)

See description of these conditions in thoracic section. Characteristics as for thoracic pain of similar etiology.

Differential Diagnosis
Also includes entrapment in rectus sheath or operative scars. Post-traumatic pain often has continuous ache with paroxysmal exacerbations.

Code
406.X2 Postinfectious
406.X8 Unknown
406.X1 or 403.X1 Post-traumatic

Twelfth Rib Syndrome (XX-4)

Definition
Chronic pain in the loin, sometimes with acute exacerbations and radiation to the groin.

Site
Eleventh or twelfth rib, or both.

Systems
Skeletal and nervous systems.

Main Features
A fairly common condition that seems to occur more often in women than men (4:1). Patients usually develop the problem between the ages of 20 and 40 years. There is not usually any history of trauma, but it may start during a pregnancy or following loin surgery. The pain may take the form of a sharp pain or a dull ache, or a combination of the two (the initial lancinating pain being followed by a prolonged period of aching pain). Patients are rarely free from pain, although the intensity varies from time to time. An attack of severe pain can be bad enough to mimic ureteric colic. The sharp pains usually last for several hours, and the subsequent dull ache subsides over a couple of days.

Aggravating Factors
Certain movements, involving alternating flexion and extension of the spine, e.g., using a vacuum cleaner.

Relief
Flexion of the spine (i.e., sitting forward).

Signs
Tenderness of the affected ribs. Manipulation of the ribs should exactly reproduce the patient's syndrome.

Laboratory Findings
None diagnostic but a chest X-ray, intravenous urogram, and spinal X-rays will help to exclude other causes of loin pain.

Usual Course
Pain continues indefinitely.

Complications
Depression and other psychological sequelae.

Social and Physical Disability
Quality of life moderately or severely impaired. Patients usually limit physical activities lest they provoke an acute attack.

Pathology
No histological abnormality identified in ribs. It is assumed that the cause is irritation of an intercostal nerve by the offending rib.

Summary of Essential Features and Diagnostic Criteria
Loin pain, either intermittent or continuous and sometimes with radiation to the groin. Frequently misdiagnosed as pain of renal origin. Diagnosis is clinical and depends upon exactly reproducing the patient's pain by palpation of the rib. Confirmatory evidence can often be obtained by using local anesthetic to block the appropriate intercostal nerve, but a negative test would not necessarily exclude the syndrome.

Treatment
Reassure the patient of the benign nature of the condition. Surgical exploration is a consideration.

Differential Diagnosis
Renal or ureteric pathology, spinal problems, pulmonary pathology.

Code
433.X6a

Reference
Machin DG, Shennan JM. Twelfth rib syndrome: a differential diagnosis of loin pain. Br Med J 1983;287:586.

Abdominal Cutaneous Nerve Entrapment Syndrome (XX-5)

Definition
Segmental pain in the abdominal wall due to cutaneous nerve entrapment in its muscular layers, commonly at the outer border of the rectus sheath or by involvement in postoperative scar tissue.

Site
Unilateral in the abdomen, usually confined to a single dermatome.

System
Peripheral nervous system.

Main Features
Initially there is abdominal wall pain, which is sharp and burning but intermittent. Later the patients typically complain of a constant dull ache, with an additional sharp, stabbing pain in the anterolateral subcostal region on twisting, coughing, or straining. The aching pain is worse when sitting and easier when standing or walking. Pressure of tight garments over the nerve can aggravate the pain.

With nerve entrapment in the rectus sheath the pain occurs, or is made worse, when the abdominal wall is tensed, for example if the patient is asked to raise the head and neck off the examining couch. The diagnosis is frequently missed when the abdomen is relaxed, as it is for conventional examination. The diagnosis may also be supported by the response of pain on localized pressure of the fingertip, pencil head, or similar object over the tender area.

The measures in examination assist in determining which thoracic nerve is trapped and may require injection.

Relief
Relief is obtained immediately by injection of local anesthetic into the trigger zone.

Differential Diagnosis
Serious intra-abdominal pathology, such as acute appendicitis, is normally not so prolonged over weeks or months. The pain of appendicitis is present even when the abdomen is relaxed and usually is associated with other well-known physical signs. Entrapment neuropathy may require distinction from other causes of segmental pain (see Intercostal Neuralgia).

Code
433.X6b

GROUP XXI: ABDOMINAL PAIN OF VISCERAL ORIGIN

Cardiac Failure (XXI-1)

Definition
Dull aching pain from congestive heart failure.

Site
Pain from congestive heart failure is usually epigastric or in the right upper abdominal quadrant.

System
Gastrointestinal system. Pain is thought to be related to distension of the hepatic capsule. Bowel ischemia or congestion may also be a factor.

Main Features
Dull aching pain in association with a tender enlarged liver and other signs of congestive heart failure.

Associated Symptoms
Dyspnea, increased abdominal girth, ankle edema, decreased exercise tolerance.

Signs and Laboratory Findings
Physical findings of congestive heart failure may include crackles on auscultation, elevated jugular venous pressure, hepatomegaly, and occasionally a pulsatile liver, ascites, and edema. An S3 and S4 gallop may be heard. Chest X-ray may show cardiomegaly and pulmonary edema. LDH, bilirubin, and SGOT may be elevated secondary to hepatic congestion.

Usual Course
This is variable depending on the treatability of the congestive failure. The pain may settle promptly with good medical management.

Complications
Long term this may result in “cardiac cirrhosis.”

Social and Physical Disability
If prolonged, it may be part of a disability secondary to heart failure.

Pathology
Passive congestion of the liver is the pathological finding. The primary pathology is usually coronary artery disease.

Essential Factors
Dull aching right upper quadrant and epigastric pain with a large tender liver and elevated liver enzymes in association with other findings of heart failure.

Differential Diagnosis
Hepatitis and diseases of the gallbladder.

Code
452.X6

Gallbladder Disease (XXI-2)

Definition
Pain due to an inflammatory disorder of the gallbladder usually associated with gallstones. Specific disease process must be demonstrated.

Site
Right upper quadrant, but also epigastrium and other parts of the abdomen.

System
Gastrointestinal; gallbladder and bile duct.

Main Features
Prevalence: common, especially in middle age, except in ethnic minorities with high prevalence when younger age groups are also often affected (e.g., some North American Indians). *Sex Ratio:* much more common in women. *Pain Quality:* pain associated with passage of stone into the cystic duct is a severe colic, short lived with associated sweating.

Associated Symptoms
Anorexia, nausea and vomiting, jaundice, dark urine, pale stool. Relieved with antispasmodics. Dyspepsia with fatty foods.

Signs and Laboratory Findings
Tenderness in right upper quadrant. Neutrophil leucocytosis; hyperbilirubinemia; elevation in serum transaminases and alkaline phosphatase. Evidence also from ultrasound (only rarely are cholecystograms needed).

Usual Course
Resolves within two or three days unless stone impacts in common bile duct, causing obstructive jaundice.

Complications
Obstructive jaundice, mucocele of the gallbladder, empyema of gallbladder with or without rupture.

Pathology
Gallstones may be cholesterol from lithogenic bile, pigment secondary to chronic hemolysis, or mixed.

Summary of Essential Features and Diagnostic Criteria
Acute right upper quadrant pain, dyspepsia to fatty foods. Diagnosis by ultrasound or cholecystogram showing duct dilatation.

Differential Diagnosis
Hepatitis, renal colic, hepatic flexure syndrome.

Code
456.X6

Post-cholecystectomy Syndrome (XXI-3)

Definition
Right upper quadrant pain in patients following cholecystectomy.

Site
Right upper quadrant.

System
Gastrointestinal (external biliary tree).

Main Features
Prevalence: this pain is a common occurrence soon after the gallbladder has been removed, often with a short initial pain-free period. *Sex Ratio:* it is more common in females. *Pain Quality:* the pain is similar to "gallbladder" pain, may be colicky in nature, daily, but not at night, may be dull or very intense lasting all day, and may continue for months or years.

Associated Symptoms
Nausea, occasionally vomiting. Aggravated by eating.

Signs and Laboratory Findings
Tenderness in right upper quadrant in region of the scar. No abnormal laboratory tests.

Usual Course
Chronic, unrelenting.

Complications
Risk of misuse of opioids or further unnecessary surgery.

Social and Physical Disability
Those of chronic pain.

Summary of Essential Features and Diagnostic Criteria
Right upper quadrant pain in a patient following cholecystectomy with no obvious cause. Endoscopic retrograde cholangiography often reproduces the pain.

Differential Diagnosis
Retained bile duct stone, hepatic flexure syndrome.

Code
457.X1

Chronic Gastric Ulcer (XXI-4)

Definition
Attacks of periodic upper abdominal pain due to ulceration of the gastric mucosa.

Site
Pain is generally rather diffuse over the central upper abdomen. It may radiate in any direction and occasionally through to the back.

System
Gastrointestinal system.

Main Features
Sex Ratio: males and females are about equally affected, although in some areas it is more common in females, e.g., Australia. *Age of Onset:* can occur at any age, but most common in the middle-aged and the elderly. *Time Pattern:* sudden onset of pain after meals from within one-half to two hours. There is a variable relationship to meals, typically (but not always) being aggravated by eating. Pain may be relieved by fasting or antacids. At first may be periodic and infrequent, every two to three months lasting for a few

days.

Associated Symptoms
Anorexia and mild weight loss, often nausea, but vomiting is rare and associated with a prepyloric ulcer.

Signs and Laboratory Findings
May be anemic. Patient shows site of pain by pointing to diffuse area of upper abdomen with hand. Tender on palpation in that area. The diagnosis is made on endoscopy (or rarely barium meal). Mild iron-deficiency anemia, or elevation of ESR, or both may be found on blood examination.

Usual Course
Periodic pain becomes more frequent and perhaps severe and for longer duration until pain-free periods may disappear. Pain commonly responds to regular antacid and anticholinergic therapy and particularly to H2 receptor antagonists, but there is a high incidence of relapse. Commonly associated with infection by *Helicobacter pylori,* which needs eradication to allow healing.

Complications
Gastric ulcers may bleed, usually chronically, presenting with iron-deficiency anemia but occasionally acutely presenting with hematemesis and melena; chronic ulceration leads to scarring so that prepyloric ulcers may cause obstruction with vomiting. Peptic ulcers may perforate, though usually insidiously, resulting in erosion into adjacent structures such as the pancreas. This causes localized but rarely generalized pancreatitis, or acute perforation with resulting acute peritonitis.

Social and Physical Disability
Recurrent or chronic pain will restrict normal activities and reduce productivity at work.

Pathology
Chronic ulceration with transmural inflammation results in localized fibrosis and cicatrization.

Summary of Essential Features and Diagnostic Criteria
Chronic gastric ulcer is a syndrome of periodic diffuse postprandial upper abdominal pain relieved by antacids. The diagnosis is made by endoscopy (or barium contrast radiology).

Code
455.X3a

Chronic Duodenal Ulcer (XXI-5)

Definition
Attacks of periodic epigastric pain due to ulceration of the first part of the duodenal mucosa.

Site
Pain is classically localized to a spot high in the epigastrium, either central or under the right costal margin, and commonly projects through to the back.

System
Gastrointestinal.

Main Features
Occurs at any age but commonly in young and middle-aged adults and is still more common in men. However, the incidence is less than 2:1, males to females. Commonly occurs when the patient is fasting, especially at night, and is relieved by eating or antacids. There is a variable relationship to meals, typically (but not always) being eased by eating. Periodic pain, which commonly lasts from a few days to

two or three weeks, with pain-free periods lasting for months.

Associated Symptoms
Weight loss uncommon; patients may actually gain weight. Dyspepsia and often nausea occur, but vomiting is uncommon.

Signs and Laboratory Findings
Patient often points to site of pain, which is also tender, with one finger. The diagnosis is made on endoscopy or barium meal (upper gastrointestinal series). Mild iron deficiency anemia and elevated ESR may occur. Rarely hypercalcemia is discovered in association with hyperparathyroidism.

Usual Course
Attacks of periodic pain may become more frequent and for longer duration. Pain commonly responds to appropriate doses of antacids and healing is promoted by H2 receptor antagonists. But there is a high incidence of relapse, which can be considerably prevented by maintenance doses. Commonly associated with infection by *Helicobacter pylori,* which needs eradication to allow healing.

Complications
Duodenal ulcers may acutely bleed or perforate.

Social and Physical Disability
Restriction of normal activities and reduction of productivity at work.

Pathology
Chronic ulceration with transmural inflammation resulting in localized fibrosis and cicatrization.

Summary of Essential Features and Diagnostic Criteria
Chronic duodenal ulcer is a syndrome of periodic, highly localized, upper epigastric pain relieved by antacids. The diagnosis is made by endoscopy or barium contrast radiology.

Code
455.X3b

Carcinoma of the Stomach (XXI-6)

Definition
Constant upper abdominal pain due to neoplasm of the stomach.

Site
Anywhere in the upper abdomen.

System
Gastrointestinal system.

Main Features
Uncommon, occurring predominantly in middle-aged and elderly patients but can occur in the third decade of life. There may be a past history of a gastric ulcer or partial gastrectomy 15 years or more previously. Pain varies from a dull discomfort to an ulcer-like pain, which is not relieved by antacids, to a constant dull pain.

Associated Symptoms
Anorexia and weight loss early in the disease, together with fatigue. The patient may present with acute

gastrointestinal bleeding, hematemesis and/or melena, or signs of anemia, e.g., fatigue, shortness of breath on exertion, and even angina and swelling of the ankles. Later, symptoms of obstruction either at the pylorus, with gastric distension and forceful vomiting, or at the cardia, with dysphagia and regurgitation, may occur.

Signs and Laboratory Findings
Physical findings include those of obvious weight loss of cachexia, a palpable mass in the epigastrium, and an enlarged liver. Laboratory findings are mainly of anemia, which may be microcytic due to chronic blood loss, normocytic due to chronic disease, or macrocytic due to achlorhydria and even to underlying pernicious anemia. Occult blood is commonly present in the stool. Hypoproteinemia is found, at times associated with a protein-losing enteropathy. Liver chemistry tests, especially alkaline phosphatase, will be abnormal in patients with hepatic metastases.

Usual Course
If the patient presents early in the course of the disease the tumor may be resectable, although the chance of recurrence in the local lymph glands is high.

Complications
There may be obstruction at the cardia or pylorus, or metastases in the liver or in more distant organs such as the lungs or bone, resulting in bone pain.

Social and Physical Disability
Inoperable patients continue with anorexia and weight loss, become cachectic and totally incapacitated.

Pathology
The tumor is usually an adenocarcinoma. It may present as an ulcerating lesion or with diffuse infiltration of the stomach wall (linitis plastica).

Summary of Essential Features and Diagnostic Criteria
Indefinite onset of anorexia, weight loss, and fatigue in an elderly patient with vague upper abdominal discomfort developing into constant upper abdominal pain associated with anemia. The overall prognosis depends on the stage of the tumor at the time of diagnosis, early resectable tumors having an excellent prognosis.

Differential Diagnosis
Gastric ulcer.

Code
453.X4c

Carcinoma of the Pancreas (XXI-7)

Definition
Chronic constant abdominal pain or discomfort due to neoplasia anywhere within the pancreatic gland.

Site
Central or paraumbilical or upper abdominal over the surface markings of the pancreas.

System
Gastrointestinal system.

Main Features
Uncommon, occurring predominantly in older patients, i.e., average age 65 years, but can occur in third decade of life. Pain can vary from a dull discomfort to, in the later stages, an excruciating severe pain boring through to the back, which is difficult to relieve with analgesics.

Associated Symptoms
Generalized symptoms of fatigue, anorexia, weight loss, fever, and depression occur early in the course of the disease. The patient may present with a sudden onset of diabetes mellitus late in life, without a family history, or with recurrent venous thromboses. Later symptoms include jaundice with pale stools and dark urine, pruritus, nausea, and vomiting.

Signs and Laboratory Findings
Evidence of recent weight loss and eventually cachexia are common. Jaundice and a central or lower epigastric hard mass are late findings, and a palpable spleen tip is uncommon. Laboratory findings usually show normochromic normocytic anemia with or without thrombocytosis, elevated fasting or two-hour postprandial blood glucose. Later, an elevated alkaline phosphatase and serum conjugated bilirubin may occur and the serum amylase may be slightly elevated.

Usual Course
Only a minority of patients, from 20 to 40%, are operable at the time of diagnosis. Only about 20% of those (i.e., 5 to 10% overall) have a potentially curative resection with a four-year survival of about 40%, or 4% of the whole.

Complications
These include diabetes mellitus, obstructive jaundice, portal vein thrombosis, and small or large intestinal obstruction.

Social and Physical Disability
The symptom complex with weight loss and generalized weakness is eventually totally incapacitating.

Pathology
The tumor is usually adenocarcinoma.

Summary of Essential Features and Diagnostic Criteria
Indefinite onset of anorexia, weight loss and fatigue in an elderly patient with vague central abdominal discomfort eventually turning to severe constant pain with or without obstructive jaundice. The overall prognosis even with modern imaging techniques is poor.

Differential Diagnosis
Malignancy in other organs, stricture or impacted stone in the common bile duct.

Code
453.X4b

Chronic Mesenteric Ischemia (XXI-8)

Definition
Intermittent central abdominal pain or discomfort related to ischemia of the large or small intestine.

Site

Central, periumbilical, occasionally radiating to the back.

System
Gastrointestinal system.

Main Features
Progressively severe abdominal pain precipitated by ingestion of a large meal. It may progress to almost constant pain and fear of eating.

Associated Symptoms
There may be symptoms suggestive of gastric or duodenal ulceration or intermittent incomplete small bowel obstruction.

Signs and Laboratory Findings
There may be evidence of generalized atherosclerosis as shown by absent femoral popliteal or pedal pulses, or the presence of an epigastric bruit. No specific laboratory findings are diagnostic. Weight loss is associated with a severe form of this disease. Arteriographic evaluation indicates severe stenosis or occlusion of all three mesenteric vessels, especially the superior mesenteric artery, but also possibly the inferior mesenteric artery, and the celiac axis. A meandering artery, indicating collateral blood flow to the colon, is a common finding.

Usual Course
Progressive weight loss and abdominal pain if untreated. Sudden and complete infarction of the small bowel may occur.

Social and Physical Disability
This unusual problem may be part of a picture of generalized atherosclerosis, in which case the patient may suffer from angina, cerebral vascular disease, or intermittent claudication.

Pathology
Patients with true mesenteric ischemia show severe narrowing of all three mesenteric vessels by atherosclerosis, which leads to inadequate blood flow to the gastrointestinal system. Atherosclerosis is usually isolated at the origin of these three vessels.

Summary of Essential Features and Diagnostic Criteria
Mesenteric ischemia may result in central abdominal pain, associated with ingestion of meals. When this becomes severe, weight loss results and sudden small bowel infarction may occur.

Differential Diagnosis
This rare disease is usually diagnosed by exclusion of other causes of intraabdominal pathology.

Code
455.X5

Crohn's Disease (XXI-9)

Definition
Pain due to chronic granulomatous disease of the gastrointestinal tract.

Site

Right iliac fossa if principally distal ileal involvement; generalized abdominal if colonic inflammation; less commonly anus.

System
Gastrointestinal system, sometimes including liver. Other systemic involvement is principally musculoskeletal.

Main Features
Becoming increasingly common in young adults but can occur at any age; males and females affected equally; pain usually due to intestinal smooth muscle colic, more often than obstruction in the distal ileum; localized inflammation may result in abscess formation or fistulization to other organs or skin, causing constant severe localized pain. Both pains will persist until treated.

Associated Symptoms
Intestinal obstruction associated with distention, nausea and vomiting, alteration in bowel habit, constipation or diarrhea or both, aggravated by eating, relieved by "bowel rest." Localized inflammation associated with fever, anorexia, and malaise.

Signs and Laboratory Findings
Mass in right lower quadrant; signs of previous laparotomy; evidence of fistula; central abdominal distension; increased bowel sounds if obstruction present (rare). Elevated inflammatory markers during relapse.

Usual Course
Relapsing-remitting illness most typically. The symptoms in patients with Crohn's disease will often settle with bowel rest (parenteral nutrition), with anti-inflammatory therapy (5-ASA drugs) or Metronidazole with or without corticosteroids. Refractory disease warrants consideration of monoclonal antibody therapy or resectional surgery. Extraintestinal involvement may cause joint pains.

Complications
Strictures, fistulas, nutritional failure, sacroiliitis.

Social and Physical Disability
A high proportion of patients with Crohn's disease require surgery. Fistulating disease presents major life impairment.

Etiology
Genetic predisposition with likely bacteriological triggers.

Essential Features
Pain due to a chronic inflammatory granulomatous condition of the GI tract resulting in narrowing of the ileum and inflammatory "skip" lesions of the colon. Anemia and nutritional problems are common, elevation of inflammatory markers reflect disease activity.

Differential Diagnosis
Small intestine—benign strictures; large intestine— ulcerative colitis.

Code

456.X3a	Colicky pain
452.X3a	Sustained pain

Ulcerative Colitis (XXI-10)

Definition
Pain due to mucosal inflammation of the gastrointestinal tract, and occasionally, associated smooth muscle irritability.

Site
Generalized abdominal. Colon, always the rectum and if more extensive up to the cecum.

System
Gastrointestinal system, sometimes including the liver. Other systemic involvement is principally musculoskeletal.

Main Features
Peak incidence in young adults with a smaller second peak in the 6th decade; females affected slightly more than males. Pain usually due to intestinal smooth muscle colic. In rare situations pain arises from pathological dilatation of the colon due to severe inflammation. Oral pain often due to aphthous ulceration, and limb pain may feature in associated pyoderma gangrenosum.

Associated Symptoms
Bloody diarrhea is the hallmark. Fever, anorexia, and malaise in severe disease.

Signs and Laboratory Findings
Central abdominal distension in toxic dilatation, otherwise nonspecific tenderness on palpation only. Rectal mucosa inflamed on inspection. Elevated inflammatory markers during relapse.

Usual Course
Often a relapsing-remitting illness, but can be chronically active. Symptoms in mild flare-ups will often settle with anti-inflammatory therapy (5-aminosalicylic [5-ASA] drugs). More marked inflammation may need corticosteroids. Refractory disease warrants consideration of monoclonal antibody therapy or resectional surgery: this may be followed by formation of an ileo-anal pouch or continuation with an end-ileostomy. Extraintestinal involvement may cause joint pains.

Complications
Toxic megacolon, sclerosing cholangitis, secondary joint inflammation, iritis or pyoderma gangrenosum. Increased risk of bowel cancer in extensive disease.

Social and Physical Disability
A minority of patients with ulcerative colitis require surgery. Flare-ups limit quality of life, but in between episodes this is not usually an issue.

Etiology
Likely genetic predisposition with bacteriological triggers.

Essential Features
Chronic mucosal inflammatory condition of the GI tract, always involving the rectum and possibly more proximal colon in a confluent fashion. Bloody diarrhea and pain are common, elevation of inflammatory markers reflects disease activity.

Differential Diagnosis
Crohn’s disease, infectious colitis, ischemic colitis.

Code
453X.03

Chronic Constipation (XXI-11)

Definition
Abdominal pain, usually dull, due to chronic alteration in bowel habit resulting in fewer bowel movements and diminished mean daily fecal output.

Site
Left lower quadrant and upper abdomen.

System
Gastrointestinal system.

Main Features
Affects about 10% of the population in the developed world. Common in any age group but becoming increasingly common in the elderly. More common in women during menstruation, pregnancy, and menopause. The pain is located over the cutaneous markings of the colon, most commonly in the left lower quadrant and upper abdomen over the transverse colon. The pain may vary from being constant and dull to sharp or very severe, but it never prevents sleep. It may last all day, every day, with exacerbations associated with eating; defecation may bring partial temporary relief.

Associated Symptoms
The pain may be aggravated by eating and relieved by defecation. However certain high-fiber foods such as vegetables and bulk laxatives failing to cause defecation increase the pain, as do bowel irritants. Stool softeners can relieve the pain. Abdominal bloating is almost invariable.

Signs and Laboratory Findings
The abdomen may be chronically distended; colonic fecal contents are palpable as well as the colon itself, especially the descending and transverse colon, which can be tender. The rectum may be full of hard feces (rectal constipation) or empty, but with feces palpable in the sigmoid colon on bimanual examination (sigmoid constipation). Need to exclude hypothyroidism and hypercalcemia as a cause.

Usual Course
Unless the constipation is due to some correctable abnormality, such as carcinoma or a particularly poor diet, the course is usually chronic, i.e., continuous for years.

Complications
There is a suggestion on epidemiological and experimental grounds that chronic constipation predisposes to diverticular disease and carcinoma. Neither is proven in humans. Fecal impaction, particularly in the elderly, can lead to large bowel obstruction or spurious diarrhea.

Social and Physical Disability
Severe constipation, particularly in the elderly, can cause spurious diarrhea resulting in fecal incontinence.

Pathology
Chronic constipation is most closely related to diet. The Western world's highly refined low-fiber diet predisposes to small stool weights and constipation, which is little known in Third World countries. Rarer causes include disorders of colonic muscle such as congenital megacolon and Hirschsprung's disease.

Summary of Essential Features and Diagnostic Criteria
Abdominal pain, usually dull, sometimes exacerbated by eating due to chronic constipation, which is largely a disorder of Western civilization and increases with age. The diagnosis is made from the history and physical examination.

Differential Diagnosis
Diverticular disease, carcinoma of the colon.

Code
453.X7a

Diverticular Disease of the Colon (XXI-12)

Definition
Pain, usually dull, arising in relation to multiple small sac-like projections from the lumen of the colon through the muscular wall and beyond the serosal surface.

Site
The pain is most commonly in the left lower abdominal quadrant, related to the sigmoid colon, spreading more widely if the disease involves the whole colon.

System
Gastrointestinal system.

Main Features
The pain is not a common symptom in this very common condition, which rarely presents before age 40, but becomes increasingly common with age. Males and females are equally affected. Pain may be dull and chronic, recurrent in nature, associated with constipation or acute severe pain in the left lower quadrant, associated with acute inflammation (acute diverticulosis), and lasting one to two weeks.

Associated Symptoms
Chronic constipation, acute or chronic abdominal distension, rectal bleeding (in diverticulitis). Aggravated by chronic constipation. Relieved by high cereal fiber diet (e.g., bran).

Signs and Laboratory Findings
Abdominal (colonic) distension. Palpable descending and sigmoid colons with or without tenderness. Abdominal CT scan or barium enema shows multiple diverticula.

Usual Course
Chronic disorder with constipation as a frequent problem. Acute attacks of diverticulitis occur infrequently, hallmarked by altered bowel habit, abdominal pain, rectal bleeding and fever. Patients rarely require operative intervention for subacute obstruction.

Complications
Acute diverticulitis, obstruction with or without spurious diarrhea, bleeding, peridiverticular abscess or rarely fistulization.

Social and Physical Disability
Chronic constipation and spurious diarrhea may lead to rectal incontinence. If surgery is needed, a permanent colostomy may be required.

Pathology
Hypertrophy of circular colonic muscle with penetration by sacs consisting of mucosa, connective tissue, and the serosal surface.

Summary of Essential Features and Diagnostic Criteria
A common chronic condition of the elderly resulting in constipation, colonic distension, and sometimes abdominal pain. The diagnosis is made by identification of diverticula on barium enema.

Differential Diagnosis
Chronic constipation, carcinoma of the colon.

Code
454.X6

Carcinoma of the Colon (XXI-13)

Definition
Pain due to malignant neoplasm of the large bowel.

Site
Most commonly lower abdominal or perineal pain from a lesion of the rectosigmoid area. Then any part of the abdomen from involvement of the colon, including the cecum.

System
Gastrointestinal system.

Main Features
One of the most common cancers in the developed countries, in contrast to developing countries. It is common in the middle aged and elderly. However it can occur rarely in young adults and children. The sex ratio is equal. The illness presents commonly with an alteration in bowel habit or with iron-deficiency anemia. There are several possible mechanisms of pain: the most common is due to obstruction with colon distension. Rarely pain is due to erosion through the colonic wall with peritoneal involvement. Pain is persistent and progressive until treated.

Signs and Laboratory Findings
A palpable abdominal mass or colonic distension or a palpable rectal mass. Positive fecal occult blood. Iron deficiency anemia. Visualization by barium enema or endoscopy.

Usual Course
The pain is short lived once the diagnosis is made, and it disappears with surgical removal of tumor, but pain may result later from metastases.

Complications
Acute or chronic rectal bleeding. There may be obstruction with a change in bowel habit, rarely colonic perforation or fistula formation into another viscus such as the bladder.

Social and Physical Disability
Surgical treatment may involve a permanent colostomy.

Pathology

The pathology is that of adenocarcinoma, beginning in the mucosa or in an adenomatous polyp, and spreading through the muscular wall to the serosa and via the lymphatic system and later the mesenteric blood supply to metastases to the liver, lung, etc.

Summary of Essential Features and Diagnostic Criteria
One of the most common cancers in the Western world, manifesting either as iron deficiency anemia, rectal bleeding, or an alteration in bowel habit, sometimes with abdominal or perineal pain. Diagnosed by endoscopy or barium enema.

Differential Diagnosis
Benign polyps and strictures, diverticular disease, ischemia colitis.

Code
452.X4

Gastritis and Duodenitis (XXI-14)

Code
45X.X2c

Dyspepsia and Other Disorders in the Stomach with Pain (XXI-15)

Code
45X.X7c or 45X.X8

Radiation Enterocolitis (XXI-16)

Code
453.X5

Post-Gastric Surgery Syndrome, Dumping (XXI-17)

Code
454.X1a

Chronic Pancreatitis (XXI-18)

Definition
Pain due to long-standing pancreatic inflammation and progressive destruction of the organ.

Site
Most commonly upper abdominal and back pain.

System
Gastrointestinal system.

Main Features
Persistent abdominal and back pain, severe in nature and may require opiates. Pain typically worse with

meals, especially high fat or protein ones. Steatorrhea may result from malabsorption of fatty components of diet. Severe nausea is also common. Weight loss due to malabsorption is common. Commonest causes are alcohol, gallstone disease, and auto-immune pancreatitis. It is also a complication of cystic fibrosis.

Signs and Laboratory Findings
Serum amylase and lipase may not be elevated (especially in advanced disease). Reduced fecal elastase is a more sensitive analysis. Structural confirmation by ERCP, MRCP, or CT scan is often required. Pancreatic calcification on abdominal X-ray is pathognomic, but uncommon.

Usual Course
A recurrent and often progressive condition, especially if the underlying cause is not addressed.

Complications
Diabetes is a common complication due to the chronic pancreatic damage and may require treatment with insulin. Mood disturbance is common.

Social and Physical Disability
Inappropriate use of opioids and mood disturbance are common. Mobility is often impaired due to severe back pain.

Pathology
Inflammatory infiltrate and progressive fibrosis of the gland.

Summary of Essential Features and Diagnostic Criteria
Severe chronic upper abdominal and back pain, exacerbated by meals, associated with meal-induced steatorrhea; later, and frequently, features of weight loss and malnutrition become equally prominent. Fecal elastase assay gives a functional assessment, and ERCP gives a structural one.

Differential Diagnosis
Pancreatic cancer.

Code
453.XXd

Carcinoma of the Liver or Biliary System (XXI-19)

Code
453.X4

Carcinoma of the Kidney (XXI-20)

Code
453.X4

Recurrent Abdominal Pain in Children (XXI-21)

Definition
Recurrent abdominal pain is a syndrome consisting of paroxysmal episodes of unexplained abdominal pain in children.

Site
Abdomen.

System
Probably gastrointestinal system in most cases.

Main Features
Very common, affecting 10% of children, increasing from age 4. Pain usually lasts less than one hour and is self-limiting. The child is healthy between bouts of pain.

Associated Symptoms
May be associated with nausea, vomiting, pallor, limb pains, and headache.

Signs
The pain is often periumbilical but can be anywhere in abdomen. If pain always occurs at a site other than periumbilical, the possibility of other organ system pathology (e.g., genitourinary) arises.

Laboratory Findings
Blood count, urinalysis, ESR normal.

Usual Course
Variable; in more severe cases the condition is chronic over many years.

Social and Physical Disability
Usually only during pain episodes. Disability depends on the reaction of the family, child, and doctor.

Relief
Fiber supplement, behavior management if clearly of behavioral origin.

Pathology
None.

Diagnostic Criteria
Paroxysmal abdominal pain interfering with normal activities occurring at least three times over at least three months. No organic findings.

Treatment
Cognitive-behavioral therapy for abdominal pain in children appears to be effective.

Code
456.X7

References
Apley J. The child with abdominal pains, 2nd ed. London: Blackwell Scientific; 1975.

Eccleston C, Palermo T, Williams AC, Lewandoski A, Morley S. Psychological therapies for the management of chronic and recurrent pain in children and adolescents. Cochrane Database Syst Rev 2009;CD003968.

GROUP XXII: ABDOMINAL PAIN SYNDROMES OF GENERALIZED DISEASES

Familial Mediterranean Fever (FMF) (XXII-1)

Definition
Disease of unknown cause predominant in those of Mediterranean stock, notably Sephardic Jews, Armenians, and Arabs. Classic features are periodic acute self-limiting febrile episodes with peritonitis, pleuritis, synovitis, and/or erythema resembling erysipelas.

Site
Abdomen or chest.

System
Peritoneal and pleural cavity.

Main Features
Prevalence: unknown. *Sex Ratio:* affects either sex. *Age of Onset:* attacks usually appear before age 20. Hereditary; transmitted as a single genetic characteristic with autosomal recessive inheritance. *Onset:* abdominal pain (peritoneal) most frequent presenting feature, varies in severity from mild abdominal discomfort with mild pyrexia to board-like rigidity, absent peristalsis and vomiting. *Time Pattern:* attacks settle within 48 hours, leaving no residual signs. Pleural attacks resemble peritoneal ones but are less common and usually precede or follow abdominal pain. Chest wall tenderness may be marked during attack, and transient pleural effusion may occur. Attacks occur with varying frequency. Importance is in the need to avoid unnecessary laparotomy.

Associated Symptoms
Erysipelas-like erythema over the cutaneous aspects of thighs, legs, or dorsa of feet. Arthralgias or acute arthritis involving mainly large joints such as knees or ankles. Attacks typically accompanied by fever and sometimes myalgia. Precipitants such as exercise, emotional stress, menstruation, fatty food, and cold exposure have been implicated. Relief obtained only from strong analgesics, though colchicine may diminish frequency of attacks.

Laboratory Findings
Hemocytosis may occur. Pleural fluid contains polymorphs but is sterile. Elevated CRP and leucocytosis are common.

Complications
Amyloidosis is the commonest cause of death and is chiefly nephropathic. Its occurrence is highly variable depending on race and geography. When it does occur death is usually before age 40.

Social and Physical Disability
Interruption of work when severe. No physical disability if amyloidosis does not supervene.

Pathology
Unknown.

Treatment
Colchicine is effective.

Diagnostic Criteria

Periodic attacks of peritonitis (rarely pleuritis) occurring in people chiefly of Mediterranean stock. Self-limiting and associated with fever, leucocytosis, and occasional rash. Arthralgic amyloidosis may supervene and lead to death in renal failure. Sporadic cases in people of other races have been described.

Differential Diagnosis
Other causes of peritonitis, peptic ulcer, porphyria.

Code
434.X0b or 334.X0b

Abdominal Migraine (XXII-2)

Definition
Characterized by recurrent attacks of abdominal pain, and/or vomiting occurring in association with typical migraine or as a replacement or migraine equivalent.

System
Unknown; vasospasm in the autonomic diencephalic centers has been postulated.

Site
Abdomen.

Main Features
Prevalence: unknown; but uncommon in contrast to common or classical migraine. *Sex Ratio:* males more than females. *Age of Onset:* most common in children between 2 and 11; occurs in young adults. *Aura:* prodromal symptoms may occur such as listlessness, mood disturbance, yawning or, rarely, typical aura of common migraine. *Pain:* may be anywhere in abdomen but usually epigastric or periumbilical; a diffuse burning or aching increasing in severity lasting several hours but terminated by sleep; frequently associated with nausea and vomiting and is commonly replaced by vomiting alone. *Frequency:* more common in childhood as "bilious" attacks. Attacks occur often during episodes of stress, frustration, or personal conflict.

Signs
Skin may show vasodilation; nonspecific fever has been recorded.

Laboratory Findings
EEG during attack may show mild generalized dysrhythmia, with high-voltage slow waves, thought to indicate cerebral hypoxia; transient leucocytosis may occur at height of attack.

Course
Tends to become less frequent with age and usually disappears when personal conflicts resolve.

Complications
None.

Social and Physical Disabilities
Reduced work performance in some.

Pathology
Unknown.

Summary of Essential Features and Diagnostic Criteria
Recurrent attacks of vomiting and/or abdominal pain occurring either as a migraine equivalent or associated with a migraine attack; more frequent in childhood and often associated with stress or personal conflict.

Differential Diagnosis
Gallstones; peptic ulcer, porphyria, irritable gut syndrome, etc.

Code
404.X7a

Porphyria—Hepatic Porphyrias

A group of disorders characterized by increased formation of porphyrins and/or porphyrin precursors in the liver. The principal clinical manifestations are photosensitivity and neurological lesions, which result in abdominal pain, peripheral neuropathy, and mental disturbance.

Within this group three diseases are recognized: (1) intermittent acute porphyria (IAP); (2) hereditary coproporphyria (HCP); and (3) variegate porphyria (VP).

Intermittent Acute Porphyria (IAP) (XXII-3)

Definition
Inherited disturbance of porphyrin metabolism not associated with photosensitivity, with attacks of abdominal pain as a constant feature, and sometimes variable hypertension, peripheral and central neuropathy (mainly motor), or psychosis.

Site
Abdomen, either generalized or localized.

System
Autonomic peripheral or central nervous system.

Main Features
Prevalence: exact incidence unknown. *Sex Ratio:* females to males 3:2. *Age of Onset:* after puberty. Inheritance: transmitted by a single autosomal-dominant gene with variable penetrance; positive family history commonly obtained. *Manifestations:* colicky abdominal pain, moderate or severe, generalized or localized is usually the first and most prominent syndrome. Constipation, abdominal distension, and profuse vomiting common; attacks are intermittent, lasting several days to several months with periods of remission during which symptoms are slight or absent. Attacks may be precipitated by (a) a wide variety of drugs, hormones; or (b) metabolic and nutritional factors (dieting, low carbohydrate intake).

Associated Symptoms
Neurological symptoms and signs are variable but may include peripheral neuritis (motor), autonomic, brain stem, cranial nerve, and cerebral dysfunction. Hypertension is frequent.

Signs
The abdomen is soft, tenderness is marked, and rebound tenderness is absent. Abdominal distension, slight fever, and leucocytosis may occur.

Laboratory Findings
X-rays often show areas of intestinal distension proximal to areas of spasm. Hyponatremia may be severe. Porphobilinogen and delta-aminolevulinic acid (ALA) in urine.

Usual Course
Severe cases may terminate in death from respiratory failure or from azotemia. Many, however, are clinically mild or latent and may exhibit only minor or vague complaints.

Social and Physical Disabilities
Pain often results in frequent admissions to hospital. Great caution needed when administering any drug.

Pathogenesis
The primary genetic defect is a generalized deficiency of enzyme uroporphyrinogen I synthetase acting in the pathway of heme synthesis, predominantly in the liver. This leads to depression of ALA synthetase activity and overproduction of ALA and porphobilinogen.

Essential Features
Acute intermittent abdominal colic without photosensitivity, with or without neuropsychiatric associated symptoms and hypertension, and typical urinary findings (q.v.).

Diagnostic Criteria
Intermittent abdominal pain with excess porphobilinogen and/or ALA in urine.

Differential Diagnosis
Peptic ulcer, gallstones, appendicitis, diverticulitis, irritable colon, lead poisoning, etc.

Code
404.X5a

Hereditary Coproporphyria (HCP) (XXII-4)

Definition
An inherited disturbance of porphyrin metabolism characterized by attacks of abdominal pain, occasional photosensitivity, neurological and mental disturbance (see Intermittent Acute Porphyria [IAP] [XXII-3]).

Site
Abdomen (see IAP).

System
Autonomic nervous system.

Main Features
Very rare; only a few families described; autosomal dominant; both sexes affected; often clinically silent (see IAP). Similar but milder disturbance; acute attacks often precipitated by drugs.

Associated Features
As in IAP but photosensitivity occurs, though uncommonly.

Usual Course

As in IAP but milder.

Pathology
Due to probable partial block in conversion of coproporphyrin III to protoporphyrinogen IX. Coproporphyrinogen oxidase activity decreased, probably mainly in liver.

Code
404.X5b

Variegate Porphyria (VP) (XXII-5)

(South African Genetic Porphyria or Protocoproporphyria Hereditaria)

Definition
A rare hereditary disorder of porphyria metabolism characterized by acute attacks of abdominal pain, neuropsychiatric manifestations, and photocutaneous lesions.

Site
Abdomen, diffuse or localized.

System
Autonomic nervous system.

Main Features
Prevalence: unknown. Sporadic families reported throughout the world. First reported in Dutch descendants in South Africa where incidence is 3 in 1000 Afrikaners. Autosomal dominant in either sex. *Onset:* usually in third decade, with cutaneous photosensitivity being initial feature. Attacks of abdominal pain, identical to those described in IAP (see XXII-3). Frequency: variable. Provoked by a variety of drugs, particularly barbiturates and sulfonamide, hormones, anesthetics, ethanol.

Signs
See IAP.

Laboratory Findings
Excretion of large amounts of protoporphyrin and coproporphyrin in feces. Urinary porphyrin precursors only modestly increased or normal, except during acute attack. Dehydration may lead to azotemia, and hyponatremia is common.

Usual Course
Variable; not as severe as IAP. Permanent neuropathic change can occur.

Pathology
Partial enzyme block between protoporphyrinogen and heme is postulated. No major anatomic abnormalities at autopsy.

Diagnostic Criteria
Intermittent acute abdominal pain with prominent cutaneous photosensitivity and often neuropsychiatric manifestations.

Differential Diagnosis
See IAP.

Code
404.X5c

GROUP XXIII: CHRONIC PELVIC PAIN SYNDROMES

Chronic Pelvic Pain Syndromes

Definition of Chronic Pelvic Pain
Chronic pelvic pain is chronic or persistent pain perceived* in structures related to the pelvis of either men or women. It is often associated with negative cognitive, behavioral, sexual and emotional consequences as well as with symptoms suggestive of lower urinary tract, sexual, bowel, pelvic floor or gynecological dysfunction.

In the case of documented nociceptive pain that becomes chronic/persistent through time, pain must have been continuous or recurrent for at least 6 months. That is, it can be cyclical over a six month period, such as the cyclical pain of dysmenorrhea. Six months is arbitrary; however, six months was chosen because three months was not considered long enough if we include cyclical pain conditions. If non-acute and central sensitization pain mechanisms are well documented, then the pain may be regarded as chronic, irrespective of the time period.

* Perceived indicates that the patient and clinician, to the best of their ability from the history, examination and investigations (where appropriate) has localized the pain as being perceived in the specified anatomical pelvic area.

Cyclical pain is included in our classification and hence dysmenorrhea needs to be considered as a chronic pain syndrome if it is persistent and associated with negative cognitive, behavioral, sexual, or emotional consequences.

Chronic pelvic pain may be subdivided into those conditions with well-defined classical pathology (such as infection or cancer) and those where no obvious pathology are found. For the purpose of this classification, the term “specific disease-associated pelvic pain” is proposed for the former, and “chronic pelvic pain syndrome” for the latter. The following classification only deals with chronic pelvic pain syndromes.

Definition of Chronic Pelvic Pain Syndrome
Chronic pelvic pain syndrome (CPPS) is the occurrence of chronic pelvic pain where there is no proven infection or other obvious local pathology that may account for the pain. It is often associated with negative cognitive, behavioral, sexual or emotional consequences as well as with symptoms suggestive of lower urinary tract, sexual, bowel or gynecological dysfunction. CPPS is a subdivision of chronic pelvic pain (see above).

Further Subdivision of the Chronic Pelvic Pain Syndromes
Pain perceived in the pelvis in the chronic pelvic pain syndromes may be focused within a single organ, more than one pelvic organ and even associated with systemic symptoms such as chronic fatigue syndrome, fibromyalgia or Sjögren’s syndrome.

When the pain is perceived as well localized to a single organ, some specialists may wish to consider using an end organ term such as bladder pain syndrome (see below). The use of such a phrase with the terminology “syndrome” indicates that although peripheral mechanisms may exist, central nervous system neuromodulation may be more important and systemic associations may occur. When the pain is localized to more than one organ site, the term “chronic pelvic pain syndrome” should be used. Many, including some of the authors of this text, will never subdivide by anatomy and would prefer to refer to patients with pain perceived within the pelvis and no specific disease process as suffering with chronic pelvic pain syndrome, subdivided by psychological and functional symptoms.

Psychological Considerations for Classification
Many CPPSs are associated with a range of concurrent negative psychological, behavioral, and sexual consequences that must be described and assessed. Examples are depression, anxiety, fears about pain or its implications, unhelpful coping strategies, and distress in relationships need to be considered. Both anxiety and depression can be significant important concomitants that are relevant with respect to pain, disability and a poorer quality of life. Catastrophic interpretations of pain has been shown to be a particularly salient variable, predicting patients' report of pain, disability, and poorer quality of life, over and above psychosocial variables such as depression or behavioral factors such as self-reported sexual dysfunction. It is suggested that CPPS symptoms sometimes create a sense of helplessness that can be reported as overwhelming in patients, which may be associated with the refractory nature of their symptoms. It is important to note that many of these biopsychosocial consequences are common to other persistent pain problems but may show varying degrees of salience for any one individual suffering from CPPS. In all patients with CPPS, these consequences must be clearly described as a part of the phenotype (where the term phenotype is used to indicate the observable characteristics of the syndrome).

Functional Considerations for Classification
Functional disorders, for the purpose of this document, are pathologies that have arisen secondary to changes in the control mechanisms of an organ or system. That is they are disorders characterized by disturbance of function. As an example, slow colonic transit is a functional disorder of the bowel - the normal function of the bowel is not occurring due to changes in the mechanisms that produce defecation, the bowel control is abnormal. The term is not used in the sense of a psychiatric functional disorder. Many CPPSs are associated with functional abnormalities at a local and even systemic level. These also need to be defined as a part of the phenotype.

Functional pain disorders may not express significant pathologies in the organs that appear responsible for the primary symptoms, but they are associated with substantial neurobiological, physiological and sometimes anatomical changes in the central nervous system.

Multisystem Subdivision
It is well recognized that the end organ where the pain is perceived may not be the center of pain generation. This classification is based upon the most effective accepted method of classifying and identifying different pains, i.e., by site of presentation. It is argued that keeping the end organ name in the classification is inappropriate as in most cases there are multisystem causes and effects with the result that symptoms are perceived in multiple areas. This is an area where discussions are ongoing and despite there being strong arguments for both keeping and dispensing with an end organ classification, we have not taken the umbrella approach of referring to all pains perceived in the pelvis as being described as being chronic pelvic pain syndrome.

Dyspareunia
Dyspareunia is defined as pain perceived within the pelvis associated with penetrative sex. It tells us nothing about the mechanism and may be applied to the female or male. It is usually applied to penile penetration, but is often associated with pain during insertion of any object. As well as vaginal intercourse it may apply to anal dyspareunia. It is classically subdivided into superficial and deep.

Perineal Pain Syndrome (XXIII-1)

Perineal pain syndrome is a "neuropathic-type" pain perceived in the distribution of the pudendal nerve that may be associated with symptoms and signs of rectal, urinary tract or sexual dysfunction. There is no proven obvious pathology. It is often associated with negative cognitive, behavioral, sexual and emotional consequences as well as with symptoms suggestive of lower urinary tract, sexual, bowel, or gynecological dysfunction.

This should be separated from pudendal neuralgia which is a specific disease associated pelvic pain, because the nerve is damaged.

Code
893.X8

Urological Pain Syndromes

Bladder Pain Syndrome (XXIII-2)

Bladder pain syndrome is the occurrence of persistent or recurrent pain perceived in the urinary bladder region, accompanied by at least one other symptom, such as pain worsening with bladder filling and day-time and/or night-time urinary frequency. There is no proven infection or other obvious local pathology. Bladder pain syndrome is often associated with negative cognitive, behavioral, sexual, or emotional consequences as well as with symptoms suggestive of lower urinary tract and sexual dysfunction.

Bladder pain syndrome is believed to represent a heterogeneous spectrum of disorders. There may be specific types of inflammation as a feature in subsets of patients. Localization of the pain can be difficult by examination and as a consequence another localizing symptom is required. Cystoscopy with hydrodistension and biopsy may be indicated to define phenotypes. Recently, ESSIC (International Society for the Study of Bladder Pain Syndrome) has suggested a standardized scheme of subclassifications (van de Merwe 2008 [1]) to acknowledge differences and make it easier to compare various studies.

Other terms that have been used include “interstitial cystitis”, “painful bladder syndrome”, and “PBS/IC” or “BPS/IC”; these terms are no longer recommended.

References
Krieger JN, Nyberg L Jr, Nickel JC. NIH consensus definition and classification of prostatitis. JAMA 1999;282:236–7.

van de Merwe JP, Nordling J, Bouchelouche P, Bouchelouche K, Cervigni M, Daha LK, Elneil S, Fall M, Hohlbrugger G, Irwin P, Mortensen S, van Ophoven A, Osborne JL, Peeker R, Richter B, Riedl C, Sairanen J, Tinzl M, Wyndaele JJ. Diagnostic criteria, classification, and nomenclature for painful bladder syndrome/interstitial cystitis: an ESSIC proposal. Eur Urol 2008;53:60–7.

Code
763.X8

Prostate Pain Syndrome (XXIII-3)

Prostate pain syndrome is the occurrence of persistent or recurrent episodic pain (which is convincingly reproduced by prostate palpation). There is no proven infection or other obvious local pathology. Prostate pain syndrome is often associated with negative cognitive, behavioral, sexual, or emotional consequences as well as with symptoms suggestive of lower urinary tract and sexual dysfunction.

The term “chronic prostatitis” continues to be equated with that of prostate pain syndrome. In the editors’ and others’ opinion, whereas it is recognized that this term has a long history, it is inappropriate. The National Institutes of Health (NIH) consensus (Krieger et al. 1999 [2]) includes infection (types I and II) that we feel should not be considered under prostate pain syndrome but as specific disease-associated

pelvic pain. The term prostadynia has also been used in the past but is no longer recommended by these authors. Please note that some of the authors of this document disagree with this term and suggest that chronic pelvic pain syndrome of the male is used instead of the term prostate pain syndrome agreed by the majority.

References
Krieger JN, Nyberg L, Jr., Nickel JC. NIH consensus definition and classification of prostatitis. JAMA 1999;282:236–7.

Code
863.X8

Scrotal Pain Syndrome (XXIII-4)

Scrotal pain syndrome is the occurrence of persistent or recurrent episodic pain localized within the organs of the scrotum that may be associated with symptoms suggestive of urinary tract or sexual dysfunction. There is no proven infection or other obvious local pathology. Scrotal pain syndrome is often associated with negative cognitive, behavioral, sexual, or emotional consequences as well as with symptoms suggestive of lower urinary tract and sexual dysfunction.

Scrotal pain syndrome is a generic term and is used when the site of the pain is not clearly testicular or epididymal. The pain is not in the skin of the scrotum as such, but perceived within its contents, in a similar way to idiopathic chest pain.

Code
863.X8

Testicular Pain Syndrome (XXIII-5)

Testicular pain syndrome is the occurrence of persistent or recurrent episodic pain perceived in the testis/testes and may be associated with symptoms suggestive of urinary tract or sexual dysfunction. There is no proven infection or other obvious local pathology. Testicular pain syndrome is often associated with negative cognitive, behavioral, sexual or emotional consequences as well as with symptoms suggestive of lower urinary tract and sexual dysfunction.

Previous terms have included orchitis, orchalgia, and orchiodynia. These terms are no longer recommended.

Code
863.X8

Epididymal Pain Syndrome (XXIII-6)

Epididymal pain syndrome is the occurrence of persistent or recurrent episodic pain perceived in the epididymis that may be associated with symptoms suggestive of urinary tract or sexual dysfunction. There is no proven infection or other obvious local pathology. Epididymal pain syndrome is often associated with negative cognitive, behavioral, sexual, or emotional consequences as well as with symptoms suggestive of lower urinary tract and sexual dysfunction.

Code
863.X8

Penile Pain Syndrome (XXIII-7)

Penile pain syndrome is the occurrence of pain within the penis that is not primarily in the urethra, in the absence of proven infection or other obvious local pathology. Penile pain syndrome is often associated with negative cognitive, behavioral, sexual, or emotional consequences as well as with symptoms suggestive of lower urinary tract and sexual dysfunction.

Code
863.X8

Urethral Pain Syndrome (XXIII-8)

Urethral pain syndrome is the occurrence of chronic or recurrent episodic pain perceived in the urethra, in the absence of proven infection or other obvious local pathology. Urethral pain syndrome is often associated with negative cognitive, behavioral, sexual or emotional consequences as well as with symptoms suggestive of lower urinary tract, sexual, bowel or gynecological dysfunction.

Urethral pain syndrome may occur in men and women.

Code
863.X8

Postvasectomy Scrotal Pain Syndrome (XXIII-9)

Postvasectomy scrotal pain syndrome is a scrotal pain syndrome that follows vasectomy. Postvasectomy scrotal pain syndrome is often associated with negative cognitive, behavioral, sexual, or emotional consequences as well as with symptoms suggestive of lower urinary tract and sexual dysfunction.

Postvasectomy pain may be as frequent as 1% following vasectomy, possibly more frequent. The mechanisms are poorly understood and it is, for that reason, considered a special form of scrotal pain syndrome.

References
Fall M, Baranowski AP, Elneil S, Engeler D, Hughes J, Messelink EJ, Oberpenning F, de C Williams AC; European Association of Urology. EAU guidelines on chronic pelvic pain. Eur Urol 2010;57:35–48.

Code
863.X8

Gynecological Pain Syndromes: External Genitalia

Vulvar Pain Syndrome (XXIII-10)

Vulvar pain syndrome is the occurrence of persistent or recurrent episodic vulvar pain. There is no proven infection or other local obvious pathology. It is often associated with negative cognitive, behavioral,

sexual, or emotional consequences as well as with symptoms suggestive of lower urinary tract, sexual, bowel, or gynecological dysfunction.

Although pain perceived in the vulva was subsumed under sexual disorders in the DSM-IV-R manual for classifying psychiatric disorders, there is no scientific basis for this classification and pain perceived in the vulva is best understood as a pain problem that usually has psychological consequences. There is no evidence for its classification as a psychiatric disorder.

The International Society for the Study of Vulvovaginal Disease (ISSVD) has used the term vulvodynia, where we use the term "vulvar pain syndrome". Vulvodynia, according to the ISSVD is vulvar pain that is not accounted for by any physical findings. ISSVD has defined vulvodynia as "vulvar discomfort, most often described as burning pain, occurring in the absence of relevant visible findings or a specific, clinically identifiable, neurologic disorder". If physical findings are present, the patient is said to have vulvar pain due to a specified cause. The ISSVD subdivided vulvodynia based on pain location and temporal characteristics of the pain (e.g., provoked or unprovoked). The following definitions are based on that approach.

Code
863.X8

Generalized Vulvar Pain Syndrome (XXIII-11)

Generalized vulvar pain syndrome refers to a vulvar pain syndrome where the pain/burning cannot be consistently and precisely localized by point-pressure 'mapping' via probing with a cotton-tipped applicator or similar instrument. Rather, the pain is diffuse and affects all locations of the vulva. The vulval vestibule (part of the vulva which lies between the labia minora into which the urethral meatus and vaginal introitus open) may be involved but the discomfort is not limited to the vestibule. This pain syndrome is often associated with negative cognitive, behavioral, sexual, and emotional consequences. Previous terms have included "dysesthetic vulvodynia" and "essential vulvodynia", but are no longer recommended.

Code
863.X8

Localized Vulvar Pain Syndrome (XXIII-12)

Localized vulvar pain syndrome refers to pain that can be consistently and precisely localized by point-pressure mapping to one or more portions of the vulva. Clinically, the pain usually occurs as a result of provocation (touch, pressure, or friction).

Localized vulvar pain syndrome can be subdivided into vestibular pain syndrome and clitoral pain syndrome.

VESTIBULAR PAIN SYNDROME refers to pain that can be localized by point-pressure mapping to one or more portions of the vulval vestibule. Previous terms have included vulval/vulvar vestibulitis , vestibulodynia, and localized and focal vulvitis, but are no longer recommended.

CLITORAL PAIN SYNDROME refers to pain that can be localized by point-pressure mapping to the clitoris or is well perceived in the area of the clitoris.

Code
863.X8

Gynecological System: Internal Pelvic Pain Syndromes

Endometriosis-Associated Pain Syndrome (XXIII-13)

Endometriosis-associated pain syndrome is chronic or recurrent pelvic pain in laparoscopically confirmed endometriosis and the term is used when the symptoms persist despite adequate endometriosis treatment. It is often associated with negative cognitive, behavioral, sexual, or emotional consequences as well as with symptoms suggestive of lower urinary tract, sexual, bowel or gynecological dysfunction.

Many patients have pain above and beyond the endometriotic lesions; this term is used to cover that group of patients. Endometriosis may be an incidental finding, is not always painful and the degree of disease seen laparoscopically does not correlate with severity of symptoms. As with other patients, they often have more than one end organ involved. It has been suggested that this phenotype should be removed from the classification as the endometriosis may be irrelevant.

Code
763.X8

Chronic Pelvic Pain Syndrome with Cyclical Exacerbations (XXIII-14)

This term would cover both the non-gynecological organ pains which frequently show cyclical exacerbations (e.g. IBS, bladder pain syndrome) but also pain similar to that associated with endometriosis/adenomyosis but where no pathology is identified. This condition is different from dysmenorrhoea where pain is only present with menstruation

Code
795.X8

Primary Dysmenorrhea (XXIII-15)

Primary dysmenorrhea is pain with menstruation not associated with a well-defined pathology. Dysmenorrhoea needs to be considered as a chronic pain syndrome if it is persistent and associated with negative cognitive, behavioral, sexual, or emotional consequences.

Code
765.X8

Musculoskeletal System

Pelvic Floor Muscle Pain Syndrome (XXIII-16)

Pelvic floor muscle pain syndrome is the occurrence of persistent or recurrent, episodic, pelvic floor pain.

There is no proven well defined local pathology. It is often associated with negative cognitive, behavioral, sexual or emotional consequences as well as with symptoms suggestive of lower urinary tract, sexual, bowel or gynecological dysfunction.

This syndrome may be associated with overactivity of or trigger points within the pelvic floor muscles. Trigger points may also be found in multiple muscles, such as the abdominal, thigh and paraspinal muscles and even muscles not directly related to the pelvis.

Code
833.X8

Coccyx Pain Syndrome (XXIII-17)

Coccyx pain syndrome is the occurrence of chronic or recurrent episodic pain perceived in the region of the coccyx, in the absence of proven infection or other obvious local pathology. Coccyx pain syndrome is often associated with negative cognitive, behavioral, sexual or emotional consequences as well as with symptoms suggestive of lower urinary tract, sexual, bowel or gynecological dysfunction. The term "coccydynia" was used but is no longer recommended.

Code
533.X8

Colorectal Pelvic Pain Syndromes

Irritable Bowel Syndrome (XXIII-18)

Irritable bowel syndrome is the occurrence of chronic or recurrent episodic pain perceived in the bowel, in the absence of proven infection or other obvious local pathology. Bowel dysfunction is frequent. Irritable bowel syndrome is often associated with worry and preoccupation about bowel function, negative cognitive, behavioral, sexual, or emotional consequences as well as with symptoms suggestive of lower urinary tract, or gynecological dysfunction.

The above classification is based upon the Rome III Criteria: 3 months of continuous or recurring symptoms of abdominal pain or irritation that may be relieved with a bowel movement, may be coupled with a change in frequency, or may be related to a change in the consistency of stools.

Two or more of the following are present at least 25 percent (one quarter) of the time: A change in stool frequency (more than 3 bowel movement per day or fewer than 3 bowel movements per week), noticeable difference in stool form (hard, loose and watery stools or poorly formed stools), passage of mucus in stools, bloating or feeling of abdominal distension, altered stool passage (e.g. sensations of incomplete evacuation, straining, or urgency). Extra intestinal symptoms include: nausea, fatigue, full sensation after even a small meal, vomiting.

Code
753.X7b

Chronic Anal Pain Syndrome (XXIII-19)

Chronic anal pain syndrome is the occurrence of chronic or recurrent episodic pain perceived in the anus, in the absence of proven infection or other obvious local pathology. Chronic anal pain syndrome is often associated with negative cognitive, behavioral, sexual, or emotional consequences as well as with symptoms suggestive of lower urinary tract, sexual, bowel, or gynecological dysfunction.

Code
853.X8

Intermittent Chronic Anal Pain Syndrome (XXIII-20)

Intermittent chronic anal pain syndrome refers to severe, brief, episodic pain that seems to arise in the rectum or anal canal and occurs at irregular intervals. This is unrelated to the need to or the process of defecation. It may be considered a subgroup of the chronic anal pain syndromes. It was previously known as “proctalgia fugax”; this term is no longer recommended.

Code
854.X8

GROUP XXIV: DISEASES OF THE BLADDER, UTERUS, OVARIES, TESTIS, AND PROSTATE, AND THEIR ADNEXA

Mittelschmerz (XXIV-1)

Definition
Mittelschmerz, also called midcycle pain, occurs as recurrent pain episodes around the time of ovulation.

System
Female internal genital organs.

Site
Unilateral lower abdominal pain.

Main Features
Mittelschmerz occurs in approximately 50% of women at some point. The pain occurs around the time of ovulation and can last from minutes to 48 hours.

Associated Symptoms
Increase of cervical mucorrhea.

Signs
There may be no signs, or tenderness on bimanual palpation, especially in the corresponding iliac fossa may be present.

Complications
None.

Pathology
The pathophysiology remains to be elucidated; possible suggestions include tubal, uterine or cecal spasm; increased tension in the ovary or Graafian follicle; or peritoneal irritation due to leak of blood or fluid from the follicle. However, this latter is probably unlikely as the majority of women experience the pain prior to follicular rupture (as confirmed with US) and pain is on the same side as follicular rupture in only 86% of women.

Treatment
Simple analgesics can be used to treat the acute episode. Recurrence can be prevented by hormonal treatments that inhibit ovulation.

Diagnostic Criteria
The essential feature is recurrence around the time of ovulation.

Differential Diagnosis
Severe cases with right-sided location may erroneously be taken for appendicitis. Midcycle pain can also be caused by trapped ovary/ovarian remnant syndrome, endometriosis and ovarian cyst accidents. Ectopic pregnancy can cause similar pain and could be mistaken for Mittelschmerz in women with irregular cycles.

Code
765.X7a

Secondary Dysmenorrhea (XXIV-2)

Definition
Dysmenorrhea (pain with menstruation) is a symptom and is called secondary if a pathology is found that is probably responsible for the pain. The pain should be defined by the cause. Because 40% of endometriotic lesions remain symptomless, it should be recognized that the diagnosis of endometriosis is not necessarily a pain diagnosis, and a distinction is specified accordingly. This is dealt with specifically under the title "Endometriosis," so the diagnosis should not usually be made under "Secondary Dysmenorrhea."

System
Genital system.

Site
Pain may be unilateral or bilateral, in the lower abdomen, pelvis and lower back. Radiation into the groin and thigh may occur.

Main Features
The pain is cyclical, usually commencing just prior to and ceasing by the end of menstruation. With more severe cases, pain may commence a few days prior to menstruation. The pain is usually described as cramping in nature and may be accompanied by nausea, vomiting, fainting and altered bowel habit.

Main Causes, Associated Features, and Diagnosis
The additional features associated with secondary dysmenorrhoea and its diagnosis depend on the cause. The main pathologies causing secondary dysmenorrhea are: endometriosis, adenomyosis, pelvic inflammatory disease (PID), fibroids, obstruction and intrauterine devices. Endometriosis, adenomyosis, and PID are considered in separate sections.

FIBROIDS Fibroids do not always cause pain or dysmenorrhea and thus may be an incidental finding. Those indenting the uterine cavity often cause menorrhagia, whilst particularly bulky uteri, posterior and cervical fibroids can be associated with dyspareunia. Diagnosis: a bulky uterus may be found on bimanual palpation. Both US and MRI can be used to identify and locate fibroids, whilst those indenting the uterine cavity will be visible on hysteroscopy.

OBSTRUCTIVE DYSMENORRHEA Secondary dysmenorrhea is called obstructive when obstruction of the menstrual flow occurs, this can be either congenital or acquired. Main Features: the prevalence is difficult to evaluate. In congenital forms the pain mostly begins a few months after menarche, as it starts only when enough blood has been retained to distend the vagina or the uterus. When there is an atresia of the hymen, there is dysmenorrhea with cryptomenorrhea as the menstrual blood is retained in the vagina. If there is retention of blood in one half of a double uterus, the menstrual pain will be unilateral.

Associated Symptoms: Obstructive dysmenorrhoea is usually associated with reduced or absent menstrual flow. The asymmetrical varieties of double uteri are frequently accompanied by absence or hypotrophy of one kidney. The retained blood may distend the vagina and the uterus and give rise to a retrograde menstruation, which, after a few months, may cause implantation of menstrual debris, i.e., endometriosis.

Signs: If there is atresia of the hymen, retention of blood in the vagina will manifest itself by distention of the vagina with the hymen bulging at the introitus and the posterior wall of the vagina bulging into the rectum. Retention of blood in one half of a double uterus will cause an asymmetrical enlargement of the uterus. The distended blind half of a double vagina will bulge into the other half of the vagina.

Pathology: Various congenital anomalies may cause secondary dysmenorrhea, e.g., atresia of the hymen, a rudimentary uterine horn, a double uterus one half of which does not communicate with the vagina, or a uterus duplex bicollis, one half of which opens into a blind half of a double vagina. Acquired forms may be due to adhesions in the cervical canal after cervical surgery, including treatment of cervical intraepithelial neoplasia (CIN), and transcervical procedures such as evacuation of retained products of conception, transcervical resection of the endometrium and ablation of the endometrium. Adhesions may also be situated in the lower part of the uterine cavity known as Asherman syndrome.

Diagnostic Criteria: US or MRI can identify congenital anomalies. Hysteroscopy and sometimes laparoscopy may be required to investigate the full extent of the obstruction. Radiological exploration of the renoureteral tract is also indicated. Adhesiolysis can be performed hysteroscopically, however, the adhesions may reform.

Code
765.X6a With endometriosis
765.X4 With adenomyosis or fibrosis
765.X0 With congenital obstruction
765.X6b With acquired obstruction

Endometriosis (XXIV-3)

Definition
Lower abdominal pain secondary to foci of ectopic endometrium located outside the uterus. This is not the same as endometriosis-associated pain syndrome (see above).

System
Genital system.

Site
The pain may be located in one or in both iliac fossae or over the whole lower abdomen. It frequently projects toward the sacrogluteal region in the lower back and projects down the thighs.

Main Features
Prevalence: the true prevalence is unknown as diagnosis requires a laparoscopy. *Age of Onset:* It used to be thought that endometriosis usually develops in the late twenties or in the thirties, but since more laparoscopies have been performed on younger patients it has been found rather frequently in teenagers, and children as young as 10 years old have been shown to have biopsy-proven endometriosis. *Symptoms:* The clinical presentation of endometriosis is very variable and the extent of disease seen at laparoscopy correlates poorly with the severity of symptoms. The main symptom is pain. This may be dysmenorrhea, premenstrual pain with menstrual exacerbation, or chronic pain. Depending on the location of the ectopic implants, dyspareunia, dyschezia and dysuria may also be present. Rectal bleeding and hematuria may also occur at the time of menstruation. However, endometriosis may be asymptomatic or present with infertility.

Signs
Pelvic examination may be unremarkable. Alternatively, there may be a fixed painful retroverted uterus, or tender, enlarged, adherent adnexa on one or both sides. Small, tender nodular lesions palpated either in a sacrouterine ligament or on the posterior surface of the uterus, are almost pathognomonic of endometriosis.

Usual Course
The ectopic foci remain receptive in a variable degree to the ovarian steroids, and they will undergo the same histological changes as the "eutopic" endometrium. The ectopic tissue may grow on the surface of the peritoneum or it may become buried in a fibrous capsule. The pain may start as secondary dysmenorrhea; it may later become premenstrual as well as menstrual, or may become continuous. The pain due to endometriotic foci is usually alleviated by pregnancy. At menopause, the pain usually disappears.

Complications
Infertility is a frequent complication. Subocclusion or occlusion of the small or the large intestine is possible but infrequent. Rupture of an endometriotic cyst located in an ovary may cause an acute abdominal emergency due to irritation of the peritoneum by the old blood flowing from the ruptured cyst. Adhesions commonly form secondary to rupture, exacerbating infertility.

Pathogenesis
The pathogenesis of endometriosis remains uncertain. A combination of retrograde menstruation and an altered immune environment, allowing implantation and the development of a nerve and blood supply, is currently considered to be the most likely etiology. Endometriotic deposits have been identified in extra-pelvic locations including the umbilicus, abdominal scars and the lungs. Venous and lymphatic transport of endometrial fragments has been demonstrated and seeding of the umbilicus and surgical incisions may occur during laparoscopy or laparotomy.

Diagnostic Criteria
Laparoscopy and lesion histology is the gold standard diagnostic test. MRI and US may identify ovarian endometrioma and deeply infiltrating rectosigmoid and bladder deposits, however, they cannot identify peritoneal disease. If the history and clinical examination are suggestive of endometriosis a definitive diagnosis is not required before commencing empirical treatment.

Treatment
Treatment of endometriosis can be hormonal or surgical or combined. The appropriate treatment(s) should be decided in discussion with the woman depending on her specific constellation of symptoms and current and future fertility wishes. The majority of endometriotic tissue is hormonally sensitive and hormonal suppression has been shown to reduce endometriosis-associated pain. The available hormonal treatments include the combined oral contraceptive pill (COCP), danazol, gestrinone, progestogens, levonorgestrel intrauterine system (LNG-IUS) and gonadotropin-releasing hormone (GnRH) agonists and antagonists. They all appear to be equally effective at controlling endometriosis-related pain but have differing adverse effect profiles. In general however, symptoms return over time after stopping the treatment. Complete surgical excision of disease can be performed and has been shown to significantly reduce pain scores, improve sexual function and improve quality of life. However, particularly with extensive disease, there are significant risks associated with surgery. Neither pre- nor postoperative hormonal treatment has been shown to improve outcome measures including pain scores, however, the LNG-IUS did significantly reduce the risk of recurrence of moderate-to-severe dysmenorrhea at 1 year.

Code
764.X6

Adenomyosis (XXIV-4)

Definition
Lower abdominal or pelvic pain secondary to the presence of endometrial glands and stroma within the myometrium.

System
Genital system.

Site
The pain may be located in one or in both iliac fossae or over the whole lower abdomen/pelvis. Radiation into the back and/or thighs frequently occurs.

Main Features
Adenomyosis may be a cause of chronic pelvic pain. It shares many features with endometriosis and the two conditions frequently coexist. Diagnosis was previously histological, with adenomyotic deposits identified in between 5% and 70% of hysterectomy specimens, depending on the indication for surgery. It was thought to be a disease of women in their fourties and fifties, but is increasingly diagnosed in younger women.

The main symptom is pain, often preceding the onset of menstruation and then exacerbated by menstruation. Menorrhagia, metrorrhagia and dyspareunia are frequently also present.

Signs
Pelvic examination may reveal a bulky, tender uterus.

Complications
Adenomyosis may be associated with infertility.

Pathology
Adenomyosis is thought to occur secondary to a breach in the integrity of the myometrial-endometrial junction. Thus it is associated with pregnancy, especially those complicated by abnormal placentation, and surgery (including evacuation of retained products of conception [ERPC] and cesarean section). It may also occur after blunt trauma to the abdomen. There appear to be two distinct forms of the condition; diffuse, where endometrial cells are widely distributed throughout the myometrium and focal where a discrete collection of cells are seen (an adenomyoma). Typically, the surrounding myometrium is hypertrophic and hyperplastic. As with endometriosis, the ectopic endometrial tissue is hormonally sensitive.

Diagnostic Criteria
Diagnosis is histological or radiological. Both MRI and transvaginal US can be used; however, MRI is less user-dependent. Without reliable diagnostic criteria, clinical trials of treatments are difficult, and a true prevalence is unknown.

Treatment
As with endometriosis, treatment is hormonal or surgical and depends on the woman's age and reproductive wishes. Some success has been achieved with hysteroscopic excision of discrete adenomyomas, but in the majority of cases hysterectomy will be performed. GnRH agonists, danazol, and the LNG-IUS have all been evaluated as successful hormonal treatments for adenomyosis.

No code.

Chronic Pelvic Inflammatory Disease (PID) (XXIV-5)

Definition
Pain where there may have been a history of chronic low-grade infection of the pelvis.

System
Genital system.

Main Features
Pain in the lower abdomen/pelvis, sometimes in the back. A history of an acute episode (or repeated episodes) of PID may be present, however, the initial infection may have been asymptomatic. Dyspareunia and offensive vaginal discharge may be present; however, fever is usually absent. Infertility is often associated.

Signs
Tenderness on bimanual palpation. A fixed retroverted uterus or bulky tender adnexa may also be found.

Diagnostic Criteria
Diagnosis is usually made on the history and examination. High vaginal and endocervical swabs should be obtained but are often negative. Ultrasound may demonstrate features of chronic salpingitis or a salpingo-ovarian abscess and severe adhesions may be seen at laparoscopy. Microbiological samples should be obtained at laparoscopy. Rarely a pyosalpinx is identified.

Treatment
Any pathogens identified should be treated with the appropriate antimicrobials. Adhesions can be divided laparoscopically; however, while this may improve fertility, the pain may persist.

Subgroups of Chronic PID
POSTERIOR PARAMETRITIS Main Features: this is rare but may occur after delivery with an unidentified cervical tear. Treatment is with broad spectrum antibiotics.

TUBERCULOUS SALPINGITIS Main Features: Rare in the developed world. *Symptoms:* The most frequent symptoms are sterility, pelvic pain, poor general condition, and menstrual disturbances. Genital tuberculosis presents under two forms, either the silent or the active form. In the silent forms there are no particular symptoms; there is no pain and no fever. In the active or advanced forms there are general symptoms and signs of the tuberculous process, meno- or metrorrhagias, sometimes amenorrhea. Pelvic pain may be present. In the active cases there is usually pyrexia, weight loss, and night sweats. *Signs:* on pelvic examination a fixed retroversion with palpable tubo-ovarian masses may be found. Spontaneous pain and dysmenorrhea may be explained by a pyo-or hydrosalpinx or by a tuberculous pelvioperitonitis. Dyspareunia may be due to a fixed retroversion or to adherent adnexal masses. *Usual Course:* The tuberculous process may become latent or may heal spontaneously. It may, on the other hand, evolve towards a pyosalpinx or an ovarian abscess or to a tuberculous pelvioperitonitis or a general peritonitis. *Diagnostic Criteria:* In advanced cases general symptoms and signs of the tuberculous process, abdominal pain or discomfort, signs of a pelvic infection, together with a positive tuberculin test and bacteriological evidence of tuberculosis constitute the basis of the diagnosis. Tubercle bacilli may be cultured either from menstrual blood or from an endometrial biopsy, taken preferably in the premenstrual phase. Silent cases are usually diagnosed by the presence of tubercular lesions in an endometrial biopsy taken during the evaluation of infertility cases.

Treatment
Treatment is essentially medical by means of a long lasting combined drug regimen appropriate to local sensitivity patterns. Surgery will be resorted to only if pelvic masses persist or increase under medical treatment, if endometrial lesions persist, and if pain or other pelvic symptoms are not alleviated by drug therapy.

Code
763.X2

Ovarian Pain (XXIV-6)

Definition
Lower abdominal pain due to pathology associated with the ovary.

RECURRENT PAINFUL FUNCTIONAL OVARIAN CYSTS Main Features: lower abdominal pain due to recurrent painful functional cysts is sometimes, although rarely, seen in young women. *Diagnostic Criteria:* if the condition is very painful, laparoscopy may be indicated in order to ascertain the cause of the pain; the cystic fluid may then be aspirated and submitted to cytological examination. If the result of this examination is compatible with a functional cyst, it is recommended to treat it conservatively by means of oral contraceptives. There is a good chance that the cyst and the pain will disappear, whereas surgical exploration with wedge resection of the ovary is likely to be followed by a recurrence of the cyst and of the painful episode, as well as potential damage to oocyte reserve.

Code
764.X7a

OVARIAN REMNANT SYNDROME Pain due to ovarian remnants following surgery. *Main Features:* when a bilateral oophorectomy has been performed in conditions that make it difficult to be sure that all ovarian tissue is removed, e.g., when the ovaries were embedded in endometriotic scar tissue or were surrounded by dense adhesions, active rests of ovarian tissue may cause a painful condition called the ovarian remnant syndrome. *Diagnostic Criteria:* an ovarian remnant will be suspected when the history is of cyclical pain despite previous bilateral oophorectomy. Pain can be unilateral or bilateral depending on the location of the remnant(s). A serum hormone profile can be obtained to confirm continued estrogen production, alternatively, a therapeutic trial of gonadotropin-releasing hormone (GnRH) agonist can be performed. If ovarian suppression removes the symptoms then a remnant is a likely diagnosis. *Treatment:* Treatment can be either hormonal or surgical. Hormonal treatment involves continued suppression of ovarian function with GnRH agonists possibly in combination with add-back low-dose hormone replacement therapy (HRT). Surgical treatment involves meticulous excision of the residual ovarian tissue at laparoscopy or laparotomy; however, this is associated with a relatively high risk of complications because of distorted pelvic anatomy and scarring.

Code
764.X7b

TRAPPED OVARY SYNDROME Pain due to an ovary trapped in dense adhesions following surgery. *Main Features:* Subsequent to pelvic surgery (usually hysterectomy) an ovary becomes trapped in dense adhesions. *Diagnostic Criteria:* Unilateral or bilateral cyclical pain with a history of surgery. Pain is removed by ovarian suppression using a GnRH agonist. *Treatment:* Treatment can be hormonal (GnRH agonist +/- add-back HRT) or surgical release/removal of the ovary. As with ovarian remnant syndrome, surgery is associated with a relatively high risk of complications.

Pain from Urinary Tract (XXIV-7)

Code
763.XXb or 863.XX

Pain Associated with Testicular Disease (XXIV-8)

Code
868.XX

Pain from Genitourinary Malignancies

Carcinoma of the Bladder (XXIV-9)

Code
763.X4

Carcinoma of the Prostate (XXIV-10)

Code
862.X4

Cervical Cancer (XXIV-11)

Code
76X.X4

Uterine Cancer (XXIV-12)

Code
76X.X4

Ovarian Cancer (XXIV-13)

Code
76X.X4

GROUP XXV: PAIN PERCEIVED IN THE RECTUM, PERINEUM, AND EXTERNAL GENITALIA OF NOCICEPTIVE OR NEUROPATHIC CAUSE

Neuralgia of Iliohypogastric, Ilioinguinal, Genitofemoral, or Pudendal Nerves (XXV-1)

Definition
Burning or lancinating or other pain syndrome due to injury of the respective nerve, may follow surgical intervention in the hypogastric, inguinal, loin or pelvic area.

Site
Depends on the nerve and may be inguinal and / or inner thigh or the anus, perineum and / or external genitalia.

System
Peripheral nervous system.

Main Features
The pain can occur immediately after an operation but not infrequently occurs after months or years. Sometimes there is no history of operation or trauma. The pain is burning or lancinating and projects to the area supplied by the sensory nerve. For the iliohypogastric nerve the pain projects to the midline above the pubis but also laterally to the hip region. For the ilioinguinal and the genitofemoral nerve, the pain projects from the groin into the anterior part of the labia major (or the scrotum and the root of the penis) and on the inside or the anterior surfaces of the thigh, sometimes down to the knee.

Pudendal neuralgia is a neuropathic pain perceived in the distribution of the pudendal or its branches (anus, perineum, vulva, clitoris, glans penis, posterior aspect of scrotum) nerve due to a pathology of the pudendal nerve or one or more of its branches. Bladder and bowl symptoms may be present as well as sexual dysfunction. This is a neuropathic condition associated with pelvic pain and not a chronic pelvic pain syndrome.

In these pains usually the pain is continuously present, but it can be intensified by pressure over the nerve. For the anterior pains the patient frequently adopts a posture that eases discomfort, with a slight flexure of the hip and a slight forward inclination of the trunk. In the case of pudendal neuralgia, sitting is often avoided.

Signs
On examination the pain can be triggered in a narrowly circumscribed area by pressure over the nerve. Usually, there is a tenderness along the course of the nerve. In the case of the iliohypogastric and ilioinguinal nerves pain may be reproduced by pressure from near the anterior superior iliac spine to the external genitalia; when the genitofemoral nerve is involved, the internal ring of the inguinal canal can be very painful. For the pudendal nerve tenderness may be detected in the region of the ischial spine and Alcock's canal. Clinical examination helps to separate out the neurological pain from any other local cause. As a rule, cutaneous sensibility is more or less impaired in the region innervated by the affected nerve. Usually, there is an increased threshold for touch and pin prick sensation in combination with hyperalgesia; the hypoesthesia is sometimes best demonstrated with cold stimuli. In some cases scratching the skin induces less reddening or an absence of it on the affected side as compared to the

intact side, indicating loss/reduced function of afferent C-fibers. Although motor impairment of appropriate muscles can be present, this is hard to evaluate. If the iliohypogastric nerve is damaged, the lower abdominal skin reflex may be absent. Typically, with involvement of the genital branch of the genitofemoral nerve in men, the cremaster reflex is absent on the affected side.

Usual Course
Without treatment, the pain may persist for several years without tendency to improvement. Local injections and surgical repair are a consideration. Neuropathic analgesics and a multidisciplinary approach also need to be considered.

Pathology
If the nerve was sectioned during surgical intervention, histological examination may show a neuroma. If the nerve was ligated or entrapped, there may be endoneural fibrosis.

Diagnostic Criteria
Typical pain radiation with sensory impairment and pain relief by local anesthetic.

Treatment
The pain can be relieved by injection of a local anesthetic proximally from the injury side, steroid is often included; for the iliohypogastric and ilioinguinal nerve the injection is done two finger-widths medially from the anterior superior iliac spine, where they leave the internal oblique muscle. For the pudendal nerve, injections may be at the ischial spine with x-ray guidance and/or within Alcock's canal under CT guidance. Surgical exploration, medications for neuropathic pain and a multidisciplinary approach should be considered.

Diagnostic Criteria
1 Burning pain with occasional superimposed paroxysms in the distribution of the involved nerve.
2 Increased threshold to light touch and pinprick associated with hyperalgesia.
3 Reproduction of paroxysmal pain by pressure over the neuromata at the site of nerve injury.
4 Transient pain relief from proximal local anesthetic block.

Differential Diagnosis
Inguinal and femoral hernia; lymphadenopathy; periostitis of pubic tubercle, local muscle hyperalgesia.

Code
407.X7b
407.X1 Testicular pain

Pain of Hemorrhoids (XXV-2)

Code
853.X5

Injury of External Genitalia (XXV-3)

Code
832.X1

Ulcer of Anus or Rectum (XXV-4)

Code
81 X.XX

Gender differences for chronic abdominal and pelvic pain conditions exist; the data on prevalence is often variable.

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