EAP - Exertional Rhabdomyolysis – Sickle Cell Trait

Rhabdomyolysis is a breakdown of skeletal muscle. Rhabdomyolysis is an acute syndrome of major muscle breakdown and leakage into the bloodstream of muscle contents (electrolytes, myoglobin and other proteins) as reflected by a sharp rise in serum creatine kinase (CK).


Exertional rhabdomyolysis (ER) in athletes is a preventable and potentially fatal condition. When it occurs during a supervised strength and conditioning session, it indicates a breakdown in the structure and/or application of that session and demands a careful assessment of the reasons for that breakdown. Exertion is of the length and intensity to drive blood oxygen levels low enough to produce sickling in the sickle cell trait (SCT) patient. Further exertion can lead to more ischemia (blood vessels get blocked by sickling hemoglobin, restricting blood flow and oxygen) and subsequent rhabdomyolysis; dumping myoglobin and potassium into the blood. Lactic acidosis may occur and release more potassium further impacting heart function. Myoglobin may actually plug the kidneys and further the buildup of potassium.

Sickle Cell Trait (SCT) is not a disease. It is the inheritance of one gene for normal hemoglobin (A) and one gene for sickle hemoglobin (S), giving the genotype (AS). Sickle cell trait (AS) is not sickle cell anemia (SS), in which two abnormal genes are inherited. Sickle cell anemia causes major anemia and many clinical problems, whereas sickle cell trait causes no anemia and few clinical problems. Sickle cell trait will not turn into the disease. However, it is possible to have symptoms of the disease under extreme conditions of physical stress or low oxygen levels. In some cases, athletes with the trait have expressed significant distress, collapsed and even died during rigorous exercise. It is present in athletes at all levels. Sickle cell trait is no barrier to outstanding athletic performance. There are three constant concerns with SCT athletes: 1. Gross hematuria; 2. Splenic infarction; and 3. Exertional rhabdomyolysis.

Recognition:

- Clinical signs are often non-specific
  - Muscle pain, soreness, stiffness (DOMS: Delayed Onset Muscle Soreness)
  - Core temperature is not greatly elevated
- Separating severe ER from overlapping but milder DOMS
  - Muscle pain more severe and sustained
  - Unlike normal DOMS or muscle cramps, muscles do not twinge or spasm
  - Swelling of muscles and adjacent soft tissues
  - Weak muscles, especially in hip and shoulder girdle – slumping to the ground
  - Athlete is unable to catch breath
Collapse can happen during first 30 minutes of exertion
- Limited active and passive range of motion
- Brown (“Coca-Cola”) urine color → protein myoglobin
- Diagnostic → Elevated serum creatine kinase levels “CK Levels”
  - Elevated CK levels will be 5 times the upper limit of normal

**Increased risk:**
- Athletes who try their hardest – give their all
- Sickle cell trait student-athletes
- Not isolated to sickle cell trait student-athletes only – just more at risk
  - Student-athletes with hyperthermia
  - Student-athletes with hypothermia
  - Student-athletes with crush syndromes
  - Student-athletes with infections, autoimmune and metabolic disorders
  - Student-athletes taking certain drugs or supplements
- Workouts without a progression
- Novel workouts immediately following a transitional resting period
  - Winter break – January
  - Spring break – March
  - End of school – June/summer workouts
  - Start of school – August/traditional academic year
- Workouts following other transitional resting periods
  - Injury / Illness / Altitude changes
- Irrational intense workouts intended to punish or intimidate
- Performing exercises to muscle failure
- Increasing sets and reducing time to finish i.e. proper rest between sets/reps
- Increasing amount of weight lifted as a percentage of body weight
- Trying to condition athletes into shape in a single day or very few days (N=7-10 days)
- Conducting an unduly intense workout due to a game loss or poor performance, effort
- Hot, humid environment
Athletes with a history of dehydration

Asthma athletes

Novel overexertion is the single most common cause of exertional rhabdomyolysis
  - Too much, too soon, too fast, too long

Guidelines:

1. Identify each sickle cell student-athlete to physicians, coaches and ATCs e.g. Sickle cell testing.
2. Educate each sickle cell trait positive student-athlete about sickle trait and sickle cell disease.
3. Have each sickle cell trait positive S-A sign the notification form.
4. Transition periods are particularly vulnerable times for athletes and demand careful attention to progression in volume, intensity, mode and duration of activity. Examples of transition periods:
   a. Athletes new to the program.
   b. Athletes returning after an injury or illness.
   c. Any delayed participation relative to the team schedule.
   d. Resumption of training after an academic break (e.g., winter, spring, summer).

2. All strength and conditioning workouts should be exercise-based, scientifically sound and physiologically representative of the sport and its performance requirements.

3. Conditioning programs should begin with a work-to-rest ratio of 1-to-4.

4. The first four days of transition periods should be separate-day workouts, and all workouts:
   a. Should be documented in writing.
   b. Should be intentional.
   c. Should increase progressively in the volume, intensity, mode and duration of physical activity.

5. All strength and conditioning workouts:
   a. Should be documented in writing.
   b. Should reflect the progression, technique, and intentional increase in the volume, intensity, mode and duration of the physical activity.
   c. Should be available for review by athletics department staff

Prevention tips:

- Moderation: Avoid too much, too soon, too fast, too long
- All training programs should start slowly and build gradually – progression “Acclimation period”
- Avoid high-intensity conditioning workouts after vacations or seasonal breaks
- Workouts are meant to improve fitness, skills and athletic performance. Not for punishment or building toughness
- Set the right tone. Not meant to punish or intimidate
- Encourage athletes to set their own pace
- Strength and conditioning workouts are the highest risk factor rather than sport skills, drills or competitions
- Fluids should be regularly available and frequent breaks should be scheduled
- Post a urine-color charts. Encourage athletes to report dark urine
- If one athlete develops early signs or symptoms of possible ER, evaluate all members of the team
- Design and practice an EAP for ER

**Emergency Action Plan:**

1. Check vital signs
2. Provide supplemental oxygen by face mask even at sea level
3. Cool the athlete if needed
4. If no immediate improvement then, call 9-1-1, attach AED and prepare for CPR
5. Transport to hospital quickly and instruct doctors to prepare for explosive rhabdomyolysis and grave metabolic complications
   1. Checking urine color – Normal or darkening in color
   2. IV – normal saline
   3. High oxygen administration
   4. Check CK levels – greater than 5 times the upper limit of normal
   5. Check kidney functions
   6. Checking electrolyte values
Return to play

Phase I

1. Return to activities of daily living for about 1 to 2 weeks
2. Regular monitoring by ATC and team physician
   a. Hydration
   b. Urine color
   c. Sleep patterns
3. Monitor CK levels by team physician
   a. Decreasing levels - Get below 1000 U/L
4. Transition to Phase II as soon as CK levels normalize and no issues with ADL

Phase II

1. Reintegrate into cardiovascular work outs – 5 x per week
   a. Foam rolling / Stretching
   b. Pool work outs
   c. Stationary bike
2. Regular monitoring by ATC
   a. Hydration
   b. Urine color
   c. Muscle soreness and swelling

Phase III

1. Progression of physical activity
   a. Dynamic warm-up
   b. Functional movements
   c. Resistance training with thera-bands
   d. Core training
   e. Gradually build resistance and heart rate on stationary bike
2. Regular monitoring by ATC
   a. Hydration
   b. Urine color
   c. Muscle soreness and swelling

Phase IV

1. Begin resistance training at 20-25% or 1 rep max (RM)
   a. Untimed runs (50 yards). Run at own pace.
   b. Agility runs – 70%
   c. Gradual integration into practice
2. Regular monitoring by ATC
   a. Hydration
   b. Urine color
   c. Muscle soreness and swelling
## Signs/Symptoms of Common Non-Traumatic Collapse of Athletes

<table>
<thead>
<tr>
<th>Heat Illness</th>
<th>Sickle Cell Trait</th>
<th>Asthma</th>
<th>Cardiac</th>
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</thead>
<tbody>
<tr>
<td><strong>Heat Cramps:</strong></td>
<td>-dehydration</td>
<td>-usually known by the athlete</td>
<td>-is more instantaneous and can occur without warning</td>
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<tr>
<td>-thirst</td>
<td>-will lie fairly still after collapse and muscles appear to look normal</td>
<td>-coughing, wheezing, tightness in the chest, gasping, panicky</td>
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<td>-sweating</td>
<td>-temp &lt; 103°F</td>
<td>-on hands and knees or bent over</td>
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<tr>
<td>-transient muscle cramps (rigid muscles and muscle twinges)</td>
<td>-can occur easily and collapse will occur within the first 30 min of all out activity</td>
<td>-shortness of breath during activity or during the night</td>
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<td><strong>Heat Syncope:</strong></td>
<td>-athlete will usually stop themselves saying “I can’t go on.” Then returns to activity after sufficient rest</td>
<td>-shortness of breath in the morning or when exposed to certain allergens and irritants</td>
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<tr>
<td>-associated heat cramps</td>
<td>-differs from heat cramps</td>
<td>-the athlete with an asthma attack will usually have symptoms subside after using a prescribed albuterol inhaler</td>
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<tr>
<td>-tunnel vision</td>
<td>✓ Lack of cramping</td>
<td>-Auscultate= moving little air</td>
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<tr>
<td>-fainting</td>
<td>✓ Muscle pain</td>
<td>-usually occurs after sprinting</td>
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<td><strong>Heat Exhaustion:</strong></td>
<td>-athlete will usually stop themselves saying “I can’t go on.” Then returns to activity after sufficient rest</td>
<td>-limp or seizing</td>
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<td>-cool clammy skin</td>
<td>-differs from heat cramps</td>
<td>-body temp is irrelevant</td>
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<tr>
<td>-cramps/syncope</td>
<td>✓ Lack of cramping</td>
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<tr>
<td>-profuse sweating</td>
<td>✓ Muscle pain</td>
<td></td>
<td></td>
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<tr>
<td>-decreased urine output</td>
<td>“tetany”</td>
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<tr>
<td><strong>Heat Stroke:</strong></td>
<td>-athlete will usually stop themselves saying “I can’t go on.” Then returns to activity after sufficient rest</td>
<td>-unconscious</td>
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<td>-occurs late</td>
<td>-differs from heat cramps</td>
<td>-limp or seizing</td>
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<tr>
<td>-temp &gt; 106°F</td>
<td>✓ Lack of cramping</td>
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<tr>
<td>-irrational, confused, irritable, or incoherent</td>
<td>✓ Muscle pain</td>
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<tr>
<td>-dry hot skin, flushed face</td>
<td>“tetany”</td>
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<tr>
<td>-tachycardia</td>
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<td>-may be in coma</td>
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**MEDICAL EMERGENCY WHEN THESE ATHLETES COLLAPSE**

**Rhabdomyolosis (a breakdown of skeletal tissue that can be a cause of death in the collapsed athlete) can occur as a result of extreme heat illness OR sickle cell collapse. An athlete with this condition will have brown urine**