

Harvard University Sports Medicine Department Sickle Cell Position Statement and Guidelines

Introduction

Sickle cell trait is the inheritance of one gene for sickle hemoglobin and one for normal hemoglobin. During intense or extensive exertion, the sickle hemoglobin can change the shape of red cells from round to quarter-moon, or “sickle”. This change, exertional sickling, can pose grave risk for some athletes. In the past seven years, exertional sickling has killed nine athletes, ages 12 through 19.

Research shows how and why sickle red cells can accumulate in the bloodstream during intense exercise. Sickle cells can “logjam” blood vessels and lead to collapse from ischemic rhabdomyolysis, the rapid breakdowns of muscles starved of blood. Major metabolic problems from explosive rhabdomyolysis can threaten life. Sickling can begin in 2-3 minutes of any all-out exertion-and can reach grave levels soon thereafter if the athlete continues to struggle. Heat, dehydration, altitude, and asthma can increase the risk for or worsen sickling, even when exercise is not all-out. Despite telltale features, collapse from exertional sickling in athletes is under-recognized and often misdiagnosed. Sickling collapse is a medical emergency.

We recommend confirming sickle cell trait status in all athletes’ pre-participation physical examinations. As all 50 states screen at birth, this marker is a base element of personal health information that should be made readily available to the athlete, the athlete’s parents, and the athlete’s healthcare provider, including those providers responsible for determination of medical eligibility for participation in sports. Knowledge of sickle cell trait status can be a gateway to education and simple precautions that may prevent sickling collapse and enable athletes with sickle cell trait to thrive in sport.

Background

A condition of inheritance versus race, the sickle cell gene is common in people whose origin is from areas where malaria is widespread. Over the millennia, carrying one sickle gene fended off death from malaria, leaving one in 12 African Americans (versus one in 2,000 to one in 10,000 white Americans) with sickle cell trait. The sickle cell gene is also present in those of Mediterranean, Middle Eastern, Indian, Caribbean and South and Central American ancestry; hence, the required screening of all newborns in the United States.

In sickle cell trait, strenuous exercise evokes four forces that in concert foster sickling, 1) severe hypoxemia, 2) metabolic acidosis, 3) hyperthermia in muscles, and 4) red-cell dehydration.

Sickling Collapse: Football and Other Sports

In the past four decades, exertional sickling has killed at least 15 football players. The setting and syndrome in most are similar. Sickling players may be on-field only briefly, sprinting only 800-1,600 meters, often early in the season. Sickling can also occur during repetitive running of hills or stadium steps, during intense sustained strength training, if the temp increases late in intense one-hour drills, or at the end of practice when players run “gassers.” Sickling can even occur rarely in a game, as when a running back is in constant action during a long frantic drive downfield.

Sickling collapse is not limited to football. It has occurred in both aerobic and anaerobic sports. Sickling can begin in only 2-3 minutes of sprinting or in any other all out exertion activity.

Sickling Collapse

Sickling collapse has been mistaken for cardiac collapse or heat collapse. But unlike sickling collapse, cardiac collapse tends to be “instantaneous,” has no “cramping” with it, and the athlete (with ventricular fibrillation) who hits the ground no longer walks. Unlike heat collapse, sickling collapse often occurs within the first half hour on-field, as during initial wind sprints. Core temperature is not greatly elevated.

Sickling is often confused with heat cramping but athletes who have had both syndromes know the difference, as indicated by the following distinctions:

1. Heat cramping often has a prodrome of muscle twinges; whereas sickling has none
2. The pain is different-heat cramping pain is more excruciating
3. What stops the athlete is different-heat crampers hobble to a halt with “locked-up” muscles, while sickling players slum to the ground with weak muscles
4. Physical findings are different-heat crampers writhe in pain, with muscles visibly contracted and rock-hard; whereas sicklers lie fairly still, not yelling in pain with muscles that look and feel normal
5. The response is different-sickling players caught early and treated right recover faster than players with major heat cramping

This is not to say that all athletes who sickle present exactly the same way. How they react differs, including some stoic players who just stop, saying “I can’t go on”. As the player rests, sickle red cells regain oxygen in the lungs and most then revert to normal shape, and the athlete soon feels good again and ready to continue. This self-limiting feature surely saves lives.

Precautions and Treatment

No sickle-cell athlete is ever disqualified, because simple precautions seem to suffice. For the athlete with sickle cell trait, the following guidelines should be adhered to:

1. Build up slowly in training with pace progressions, allowing longer periods of rest and recovery between repetitions.
2. Encourage participation in preseason strength and conditioning programs to enhance the preparedness of athletes for performance testing which should be sports-specific. Athletes with sickle cell trait should be excluded from participation in performance tests such as mile runs, serial sprints, etc., as several deaths have occurred from participation in this setting.
3. Cessation of activity with onset of symptoms (muscle cramping, pain, swelling, weakness, tenderness, inability to “catch breath”, fatigue).
4. If sickle-cell trait athletes can set their own pace, they seem to do fine.
5. All athletes should participate in a year-round periodized strength and condition program that is consistent with individual needs, goals, abilities and sport-specific demands. Athletes with sickle cell trait who perform repetitive high speed sprints and/or interval training that induces high levels of lactic acid should be allowed extended recovery between repetitions since this type of condition poses special risks to these athletes.
6. Ambient heat stress, dehydration, asthma, illness, and altitude predispose the athlete with sickle cell to an onset of crisis in physical exertion.
 - a. Adjust work/rest cycles for environmental heat stress
 - b. Emphasize hydration
 - c. Control asthma
 - d. No workout if an athlete with sickle trait is ill
 - e. Watch closely the athlete with sickle cell trait who is new to altitude. Modify training and have supplemental oxygen available for competitions.
7. Educate to create an environment that encourages athletes with sickle cell trait to report any symptoms immediately; any signs or symptoms such as fatigue, difficulty breathing, leg or low back pain, or leg or low back cramping in an athlete with sickle cell trait should be assumed to be sickling.

In the event of a sickling collapse, treat it as a medical emergency by doing the following:

1. Check Vital signs.
2. Administer high-flow oxygen, 15 lpm (if available), with a non-rebreather face mask.
3. Cool the athlete, if necessary.

4. If the athlete is obtunded or as vital signs decline monitor vital signs, call EMS, apply AED, transportation to nearest hospital.
5. Tell the doctors to expect explosive rhabdomyolysis and grave metabolic complications.
6. Proactively prepare by having an Emergency Action Plan and appropriate emergency equipment for all practices and competitions.

*The above information is directly from the National Athletic Trainers Association Consensus Statement: Sickle Cell Trait and the Athlete

Sickle Cell Information and Guidelines Directly related to Harvard University Athletics

Effective August 1, 2010 all athletes participating in NCAA Div I sports must have sickle cell testing performed; show proof of sickle cell testing, or sign a waiver demonstrating that they understand the importance of testing for sickle cell, decline testing, and thereby release their institution from any liability related to declining testing, prior to being eligible to participate in any intercollegiate practice and/or strength and conditioning workouts.

- This test (a blood test) is relatively easy to perform, and can provide educational information that can be helpful to the student-athlete both on and off the field. If you were born in the U.S., your testing results may be available in your medical record.
- Athletes that are sickle cell trait positive are able to participate in sports, often with no modifications whatsoever. Individuals that test positive for sickle cell trait will have a confirmatory test performed, and if positive, will be counseled on what can be done to avoid complications.
- The medical staff will be informed of a student-athletes sickle cell status in order to provide optimal care of the student-athlete during practice, competition and conditioning.
- It is not mandatory to have the sickle cell trait test.
- The recommendation of the Harvard University Sports Medicine Department is that every student-athlete be tested or show proof of prior testing for sickle cell trait.
 - This testing should be obtained at home prior to the school year.
 - Testing will be available on campus in an appointment made basis through University Health Services.
 - If you choose to waive testing, this confirms that you understand the importance of testing, have declined and release Harvard University from any liability related to declining testing. If you are under 18 years old, a parent or guardian must sign and write their name.