



Consensus Statement: Sickle Cell Trait and the Athlete

Purpose

In a recent review of non-traumatic sports deaths in high school and college athletes (1), the top four killers, in order of occurrence, were: cardiovascular conditions, hyperthermia (heatstroke), acute rhabdomyolysis tied to sickle cell trait, and asthma. Acute exertional rhabdomyolysis (explosive muscle breakdown) from sickle cell trait is the least understood of these conditions. The purpose of this Task Force is to raise awareness of this condition and provide measures to reduce the risk of exertional collapse related to sickle cell trait.

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 a 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 breakdown 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 and 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' preparticipation 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. Nearly all of the 13 deaths in college football have been at institutions that did not screen for sickle cell trait or had a lapse in precautions for it. Small numbers preclude cogent evidence to support screening. All considered, however, we believe that each institution should carefully weigh the decision to screen based on the potential to provide key clinical information and targeted education that can save lives. Irrespective of screening, the institution should educate staff, coaches, and athletes on the potentially lethal nature of this condition.

Background

A condition of inheritance versus race, the sickle 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 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 the past four decades, exertional sickling has killed at least 15 football players. In the past seven years alone, sickling has killed nine athletes: five college football players in training, two high school athletes (one a 14-year-old female basketball player), and two 12-year-old boys training for football. Of 136 sudden, non-traumatic sports deaths in high school and college athletes over a decade, seven (5%) were from exertional sickling (1).

The U. S. military tied sickle cell trait to sudden death during recruit basic training. The relative risk of exercise-related death in sickle cell trait was about 30 (2). In other words, recruits with sickle cell trait were 30 times more likely to die during basic training. The main cause of death was rhabdomyolysis – and the risk of exertional rhabdomyolysis was about 200 times greater for those with sickle cell trait (3).

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.

Evidence supports this syndrome. Military research shows that, during intense exertion and hypoxemia, sickle cells can accumulate in the blood (4). Recent research also shows that systemic dehydration worsens exertional sickling (5). Field studies in Africa suggest that sickle-trait runners are limited not in single sprints but in middle distance or altitude running (6). The pattern in American athletes is similar.

Sickling Collapse: Football and Other Sports

The first known sickling death in college football was in 1974. A defensive back from Florida ran a conditioning test on the first day of practice at altitude in Colorado. He had collapsed on the first day of practice the year before. This time, near the end of the first long sprint, at about 700 meters, he collapsed again – and died the next day. The most recent sickling death, a freshman defensive back at Rice University in the fall of 2006, is similar. He collapsed after running 16 sprints of 100 yards each – and died the next morning. The cause of death for both athletes was acute exertional rhabdomyolysis associated with sickle cell trait.

Up to 13 college football players have died after a sickling collapse. 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 tempo increases late in intense one-hour drills, or at the end of practice when players run “gassers.”
- Sickling can even occur rarely in the game, as when a running back is in constant action during a long, frantic drive downfield (7).

Sickling collapse is not limited to football. It has occurred in distance racing and has killed or nearly killed several college or high school basketball players (two were females) in training, typically during “suicide sprints” on the court, laps on a track, or a long training run.

The harder and faster athletes go, the earlier and greater the sickling, which likely explains why exertional collapse occurs “sooner” in college football players sprinting than in military recruits running longer distances. Sickling can begin in only 2-3 minutes of sprinting – or in any other all-out exertion – and sickling can quickly increase to grave levels if the stricken athlete struggles on or is urged on by the coach.

Sickling Collapse: Telltale Features

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 talks. Unlike heat collapse, sickling collapse often occurs within the first half hour on-field, as during initial windsprints. 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 slump to the ground with weak muscles;
- 4) Physical findings are different – heat crampers writhe and yell 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 (7).

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-trait 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 paced 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-trait athletes can set their own pace, they seem to do fine.
- 5) All athletes should participate in a year-round, periodized strength and conditioning 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 conditioning poses special risk to these athletes.
- 6) Ambient heat stress, dehydration, asthma, illness, and altitude predispose the athlete with sickle trait 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 (7).

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, call 911, attach an AED, start an IV, and get the athlete to the hospital fast.
- 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.

IMMEDIATE ACTION CAN SAVE LIVES

What We Can Do

Though screening is done at birth; many athletes do not know their sickle-trait status, rendering self-report in a questionnaire unreliable. Many institutions have employed screening strategies to rectify this. A recent survey of NCAA Division I-A schools found that 64% (of respondents) screen (8). The NFL Scouting Combine screens for sickle cell trait. All considered, despite no evidence-based proof yet that screening saves lives, each institution should carefully weigh the decision to screen in the absence of documented newborn screen results.

The Consensus of this Task Force is:

- 1) There is no contraindication to participation in sport for the athlete with sickle cell trait.
- 2) Red blood cells can sickle during intense exertion, blocking blood vessels and posing a grave risk for athletes with sickle cell trait.
- 3) Screening and simple precautions may prevent deaths and help athletes with sickle cell trait thrive in their sport.
- 4) Efforts to document newborn screening results should be made during the PPE.
- 5) In the absence of newborn screening results, institutions should carefully weigh the decision to screen based on the potential to provide key clinical information and targeted education that may save lives.
- 6) Irrespective of screening, institutions should educate staff, coaches, and athletes on the potentially lethal nature of this condition.
- 7) Education and precautions work best when targeted at those athletes who need it most; therefore, institutions should carefully weigh this factor in deciding whether to screen. All told, the case for screening is strong.

Glossary

Acute Ischemic rhabdomyolysis: the rapid breakdown of muscle tissue starved of blood

Acute Rhabdomyolysis: a serious and potentially fatal condition involving the breakdown of skeletal muscle fibers resulting in the release of muscle fiber contents into the circulation

Contraindication: circumstance or condition that makes participation unsafe or inappropriate

Exertional rhabdomyolysis: muscle breakdown triggered by physical activity

Exertional sickling: hemoglobin [red blood cell] sickling due to intense or sustained physical exertion

Hyperthermia: body temperature elevated above the normal range

Hypoxemia: decreased oxygen content of arterial blood

Ischemia: a deficiency of blood flow to tissue

Metabolic acidosis: a condition in which the pH of the blood is too acidic because of the production of certain types of acids

Nontraumatic: not related to a physical injury caused by an external force

Obtunded: having diminished arousal and awareness; mentally dull

Sickling collapse: the collapse of an athlete who shows features consistent with exertional sickling

Ventricular Fibrillation: a condition in which there is uncoordinated contraction of the cardiac muscle of the ventricles in the heart

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Task Force Participants

The following individuals and associations were members of the Inter-Association Task Force on Sickle Cell Trait and the Athlete. Their participation is not an endorsement of this document. For a complete list of supporting associations, please visit http://www.nata.org/statements/consensus/sct_endorsements.htm.

Co-Chairs

Scott Anderson, ATC
E. Randy Eichner, MD

At-Large Members:

Mary L. Anzalone, MD
James C. Puffer MD
Brock Schnebel, MD

American Academy of Pediatrics
Jorge Gomez, MD

American College of Sports Medicine
Michael F. Bergeron, PhD, FACSM
Don Porter, MD

American Medical Society for Sports Medicine
James Moriarity, MD

American Orthopaedic Society for Sports Medicine
James C. Walter, II, MD

American Osteopathic Academy of Sports Medicine
Jeffrey Bytomski, DO
Angela Cavanna, DO, FAOASM

Association of Black Cardiologists
B. Wayne Kong, PhD, JD

College of American Pathologists
Michael J. Dobersen, MD, PhD

Gatorade Sports Science Institute
Jeff Kearney
Craig Horswill, PhD
Magie Lacambra, MEd, ATC

Military Medicine
Fred Brennan, Jr., DO

National Association of Basketball Coaches
Reggie Minton

National Association of EMTs
Connie Meyer, MICT

National Association of Medical Examiners
Jeffery Barnard, MD

National Athletic Trainers' Association

Veronica Ampey, MS, ATC
Douglas Casa, PhD, ATC, FACSM
Terry Dewitt, PhD, ATC
Scott Galloway, ATC, LAT
Chris A. Gillespie, MEd, ATC, LAT
Eric Howard, EdD, MS, ATC
Bob Toth, MS, ATC
Torrance Williams, ATC, LAT

National Basketball Athletic Trainers' Association
Dionne Calhoun, ATC

National Collegiate Athletics Association

David Klossner, PhD, ATC
John W. Scott, PhD, MD
Tracy Ray, MD

National Federation of State High School Associations
Bob Colgate

National Football League

Gary W. Dorshimer, MD, FACP

National Strength and Conditioning Association

Avery Faigenbaum, EdD, CSCS

Professional Football Athletic Trainers' Society

Corey Oshikoya, ATC

The Sickle Cell Disease Association of America, Inc.

National Medical Association

Betty S. Pace, MD

The Sickle Cell Foundation of Georgia, Inc.

Rudolph Jackson, MD

Women's Basketball Coaches Association

Marsha Sharp