Sickle Cell Considerations in Athletes

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KEYWORDS

• Sickle cell trait • Sudden death • Rhabdomyolysis
• Splenic infarction • Hematuria

The most vital sickle cell considerations in athletes relate to sickle cell trait (SCT). How SCT can affect athletes is replete with recent advances, updates, and controversies. The key clinical concerns for athletes with SCT are listed in **Box 1**.

These concerns are discussed in this article, along with other clinical concerns for athletes with sickling hemoglobinopathies such as hemoglobin SC, S-beta-thalassemia, and hemoglobin SE disease. Although some children and adolescents with sickle cell anemia (hemoglobin SS) compete in recreational and even team sports, secondary to anemia and cumulative complications from disease, very few compete in varsity sports in high school.[1] Therefore, sickle cell anemia is not addressed in this article.

DEFINITION, FREQUENCY, AND VARIATION OF SICKLE CELL TRAIT

SCT is not a disease but a condition, resulting from inheritance of one gene for sickle hemoglobin (S) and one gene for normal hemoglobin (A). The sickle gene is common in regions endemic with malaria, because SCT protects against early death from malaria, providing a procreation advantage to SCT carriers. SCT is found in about 8% of African American, 0.5% of Hispanic, and 0.2% of white individuals.[2] Each red blood cell in SCT typically has about 40% hemoglobin S. The co-inheritance of alpha-thalassemia trait, which occurs in about one-third of blacks (about 30% have a 1-gene and 2% have a 2-gene deletion of the 4 alpha-globin genes), lowers the amount of hemoglobin S in each red blood cell and may lessen the risk of exertional sickling.[3]

EXERTIONAL SICKLING IN ATHLETES

The most vital clinical consideration for athletes with SCT is exertional sickling, because this "sudden-collapse" syndrome can be fatal. Despite increasing national

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Box 1
Clinical concerns among athletes with SCT

1. Exertional sickling collapse
2. Lumbar paraspinal myonecrosis and other compartment syndromes
3. Splenic infarction at altitude
4. Gross hematuria
5. Hyposthenuria
6. Venous thromboembolism (questionable)

focus on this problem, fatal cases continue to occur, as illustrated by the demise of a 20-year-old male student-athlete in August 2010 during a track tryout at a university in North Carolina.[4] Other sickling collapses and deaths have occurred in various sports or exercises (**Box 2**), and have included male and female athletes, some as young as 12 years old.[5–7] Litigation over exertional sickling deaths of college football players has led to decisions by the National Collegiate Athletic Association (NCAA) on screening for SCT in college athletes. To date, the debate continues on the wisdom of this mandatory screening program and on how best to prevent tragic exertional sickling deaths.[2]

Deaths from exertional sickling have been more common in football players. The first reported case was the death of a fullback in 1963 during preseason training at a university in New Mexico. Few details are available on his death.[8] The first exertional sickling death in college football reported with some description and clinical information was in 1974.[9] A 19-year-old African American with SCT, who was a defensive back and punt returner from Florida, collapsed 2 years in a row on the first day of practice at an altitude of 5400 ft in Colorado. He survived the first year. During the second year, while aiming to finish the first conditioning sprint (880 yards), he fell behind his group at 660 yards, staggered forward, and fell at the edge of the track. He complained of severe leg pain and died the following day while hospitalized with "severe acidosis" and "severe sickling in the kidneys." In addition to these early deaths, at least 20 other deaths have occurred secondary to exertional sickling in college football players. Similar deaths have also occurred in high school and youth league football.[10]

Exertional sickling has become the leading "killer" in NCAA Division-1 football and a recent spate of deaths from exertional sickling in NCAA Division-1 football is illustrative and alarming. An examination of all deaths in Division-1 football during the past decade (2000–2010) revealed zero deaths from "play" of the game, zero deaths from "practice" of the game, and16 deaths from *conditioning* for the game, including 15

Box 2
Sports settings for exertional sickling collapse

1. Football conditioning
2. Basketball training
3. Cross-country racing
4. University track tryout
5. Golden Gloves boxing bout
6. Recreational ocean swimming