American Academy of Pediatrics
DEDICATED TO THE HEALTH OF ALL CHILDREN™
Ohio Chapter

Sports Shorts

GUIDELINES FOR PHYSICIANS

Sickle cell trait and sports

Sickle cell disease refers to a spectrum of inherited conditions (including sickle cell anemia and sickle cell trait among others) that result in the production of abnormal hemoglobin, the oxygen carrying component of red blood cells. Sickle cell disease occurs when one or both of the genes for normal hemoglobin (Hb A) are replaced by sickle hemoglobin (Hb S). Individuals with sickle cell anemia are homozygous for the sickle hemoglobin gene (genotype Hb SS) while those with sickle cell trait are heterozygous (genotype Hb AS). Sickle hemoglobin is so named because it causes the red blood cells to change shape or "sickle" which impairs their ability to transport oxygen and leads to a variety of medical problems.

How common is sickle cell trait?

In the United States, sickle cell trait is most common in those of African, Middle Eastern and Mediterranean descent because of its protective effect against malaria. Sickle cell trait occurs in about 8% of the African-American population and between 0.01-0.05% of non-black Americans.

How does sickle cell trait affect athletes?

While kids with sickle cell trait generally suffer far less morbidity than those with sickle cell anemia, exercise can induce sickling in those with trait especially if the exercise is strenuous or occurs at altitude. There are 4 major factors that contribute to sickling during exercise: hypoxemia, hyperthermia, metabolic acidosis and dehydration of the red blood cells. Once sickled, red blood cells can occlude small vessels causing tissue ischemia. This can lead to problems such as splenic infarct, exertional rhabdomyolysis and sickle collapse. Rarely, sudden death may occur from severe rhabdomyolysis-induced renal failure, electrolyte disturbance and cardiac arrhythmia.

Can sickling complications be prevented?

It is important that athletes know the symptoms and take precautions to minimize complications. However, not all athletes are aware that they have this condition. All states now test for sickle cell disease during the newborn screen and physicians should obtain these records and discuss them with the athlete at their pre-participation physical examination. If records are unavailable and status is unknown, testing should be considered at that time. Some groups, including the American Society of Hematology, believe that universal screening is unwarranted because of a lack of scientific evidence that it will save lives and a fear that testing may unfairly stigmatize those with the disease. Instead, they prefer the implementation of precautionary measures that can be utilized for all athletes regardless of sickle cell status. Precautions include: participation in off-season conditioning programs; gradually increasing training load; allowing athletes to set their own pace; and stopping activity at the onset of symptoms.

Most common symptoms of sickling

- Shortness of breath
- Fatigue
- Muscle pain or weakness (most commonly legs, buttocks and low back)
- The feeling that the athlete simply can't go any further

How should sickling in the athlete be treated?

- Stop activity
- Check vital signs
- Give oxygen
- Cool the athlete, if necessary

If improvement is not noted immediately or athlete is unresponsive:

- Call 911
- Start CPR including using an AED if available
- Get the athlete to the hospital as quickly as possible

How prevalent are these complications?

It is hard to know for sure as there is limited data. Sickle cell trait has been shown to increase the risk of exercise-related death about 30-fold in military recruits, and been linked to at least 15 deaths in college football players as well as a handful of junior and high school athletes. Other complications such as exertional rhabdomyolysis and sickle collapse may be confused with muscle cramping or heat illness and therefore are even harder to estimate.

When are complications most likely to occur?

The harder and faster athletes run, the more problems they can have. Sickling tends to increase with longer distances or with successive bursts of activity without an adequate rest period such as during repeated sprints or sustained weightlifting. Symptoms have more commonly been reported during practices than games, especially during the first few days of pre-season conditioning in the summer when the heat and humidity are high and athletes are deconditioned. Participation in sports at high altitude also significantly increases the risk of sickling.

This information is available on the Ohio AAP website www.ohioaap.org

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