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THE PEOPLE'S CLINIC: SICKLE CELL DISEASE AND ATHLETES: WHAT YOU SHOULD KNOW, WEEK OF JANUARY 27-FEBRUARY 2, 2011

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Sickle cell disease is a common blood disorder that disproportionately affects African-Americans. Approximately one in 8 African Americans carry one copy of the gene (known as sickle cell trait) and about one in 500 carry two copies of the gene (called sickle cell disease.) In sickle cell disease, the alteration in hemoglobin makes the red blood cells susceptible to becoming rigid and stiff, especially when oxygen levels are low. This shape change gives the red cells, which are normally round and smooth, a sickled shape, and these stiff, misshapen cells can clog blood vessels, especially tiny vessels. The changes in shape can be triggered by infection, fatigue, and over-exertion, changes in altitude, dehydration, low oxygen levels, and changes in body temperature. The clogged blood vessels can cause damage to organs, promote life-threatening infections, and result in severe painful episodes.

In general, persons with sickle cell trait (one copy of the gene) are relatively asymptomatic, and have a normal lifespan without any complications. Most are found to have the gene as part of mandatory testing of all newborns. However, they are more likely than persons without sickle cell trait to develop rare conditions such as blood in their urine (hematuria) or rapid breakdown of muscle fibers during extreme exertion (know as rhabdomyolysis). They are at increased risk of exertional heat illness and even exertional related death. In contrast, persons with sickle cell disease (two copies of the gene) typically have a much more complicated course, and are prone to infections, painful episodes, stroke, and lung, kidney, and heart complications, which can reduce their life expectancy to middle age or even younger.

What does this mean for athletes with the Sickle Cell trait or disease?

Conditions faced by most athletes, including vigorous exercise, dehydration, and heat can trigger the shape changes in red blood cells and result in occlusive painful episodes. Dehydration and water losses are obviously more likely to occur when exercising outdoors in hot weather, but can occur at any time, indoors or outdoors.

In general, it is not advised that persons with sickle cell disease (those that carry two copies of a sickle gene) not participate in any intense physical activity that would put them at risk of developing dehydration or vaso-occlusion. They should also avoid contact sports, such as football, in which abdominal impact could lead to rupture of an enlarged spleen. Sports should be limited to recreational level, not a competitive level. On the other hand, regular low-intensity exercise and low impact activities, such as walking, swimming, water exercise, and golf should be encouraged in persons with sickle cell disease, with close attention to such symptoms as muscle cramping, dehydration, or pain. The importance of maintaining good hydration every day cannot be overemphasized. We advise our patients to have a bottle of water with them at all times, and to drink more in the hot summer months, if they are ill, or have diarrhea. Persons with sickle cell disease should drink enough water that they need to empty their bladders at least every one to two hours throughout the day.

In contrast, persons with sickle trait can engage in physical activity and sports, as simple precautions to avoid overexertion are sufficient to prevent complications. The National Athletic Trainers' Association (NATA) published a consensus statement in 2007 to raise awareness of acute exertional rhabdomyolysis (the rapid breakdown of muscle fibers) and to reduce the risk of sickle trait related exertional deaths.

♦The athlete should be allowed to pace himself or herself, with careful attention to muscle cramping, shortness of breath back pain, chest pain, or fatigue.

♦NATA emphasizes that the harder and faster athletes go, the sooner they develop exertional collapse. Sickling can occur within with first few minutes of a sprint, or later after running longer distances.

♦Persons with sickle cell trait should not push themselves further if such symptoms occur,

nor should they be encouraged by coaches or teammates to "tough it out" or "push through" the symptoms. They should rest immediately if these symptoms occur. If collapse occurs, call 911, administer oxygen if available, and cool the athlete if in a warm environment.

◆ The athlete should be encouraged to maintain fitness year round, and to build up training slowly, with adequate rest and recovery times between repetitions.

◆ Performance tests such as mile runs, serial sprints, "suicide" sprints, should be avoided.

◆ If an athlete is ill or has an asthma exacerbation, workouts should be avoided.

◆ Changes in altitude can precipitate sickling, so athletes with sickle cell trait should be monitored closely and have their activity modified until they have acclimated to a higher altitude.

As of August 1, 2010, all new students joining NCAA National Collegiate Athletic Association Division I teams must be tested for sickle cell trait.

This mandate came in response to a lawsuit brought by the family of Dale Lloyd, a 19 year old freshman at Rice University who died after an intense football practice in 2006 and who was subsequently found to have sickle cell trait. The family hopes that the testing will prevent similar deaths in other young athletes. However, this mandate has stirred controversy regarding the implications of such testing. Though the goal of the mandate is to identify athletes who should be observed closely for signs of dehydration and excess exertion, and no athletes are to be excluded from participation on the basis of testing, it is a concern that a stigma will be created against some athletes that will outweigh the medical risk of athletes with sickle cell trait.

Thus, the precautions that should be taken by athletes with sickle cell trait are really no different than should be taken by all athletes. Maintain hydration, build up to a competitive level slowly and sensibly, and pay attention to symptom of fatigue, muscle cramping, or shortness of breath.

Coaches, parents, and trainers should establish a culture in which athletes are comfortable in reporting their symptoms and stopping to rest, without feeling pressured to push beyond these warning signs.

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