THE OFFICIAL NEWSMAGAZINE OF THE AMERICAN ACADEMY OF PEDIATRICS
AAP News
DEDICATED TO THE HEALTH OF ALL CHILDREN
Volume 33 • Number 9
September 2012
www.aapnews.org

Focus On Subspecialties

Sickle cell trait only one condition that can increase risk of sudden death during exercise

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As a result of a wrongful death lawsuit, the National Collegiate Athletic Association (NCAA) implemented a policy in August 2010 of mandatory screening of Division 1 athletes for sickle trait. Student athletes can opt out of screening if they demonstrate a past result or sign a liability waiver.

In anticipation of this policy, Jeffrey D. Hord, M.D., FAAP, and Stephen G. Rice, M.D., Ph.D., M.P.H., FAAP, covered this topic in the October 2009 issue of *AAP News*, http://aapnews.aappublications.org/content/30/10/1.2.full.pdf+html. While their recommendations remain valid, further discussion is worthwhile due to continuing controversy regarding the NCAA policy and recent recommendations in the AAP policy statement, *Climatic Heat Stress and Exercising Children and Adolescents,* http://pediatrics.aappublications.org/content/128/3/e741.full.pdf.

The AAP policy identifies numerous medical conditions that can increase a child's or adolescent's susceptibility to heat-related illness, and promotes early recognition of signs and symptoms. Perhaps most importantly, the statement focuses on appropriate preventive measures. Pediatricians must be prepared to answer questions from students and the athletic community regarding risk factors and participation.

Pediatricians should be prepared to counsel students with sickle cell trait regarding their risk for sudden death during exercise. The AAP policy *Climatic Heat Stress and Exercising Children and Adolescents* offers recommendations for all high-risk individuals.

Risk of unexplained death

A roughly 30-fold increase in unexplained deaths among military recruits with sickle trait undergoing basic training compared to black Army recruits without sickle trait was first documented in 1987 (Kark JA, et al. *N Engl J Med.* 1987;317:781-787).

Of 273 deaths among NCAA athletes from 2004 to 2008, including almost 2 million participant years, only five were attributed to sickle trait. All of these were within a group of 20 football players who died from non-traumatic medical causes. These data led to the oft-quoted 37-fold estimated increased risk of medical death in NCAA football players with sickle trait (Harmon KG, et al. *Circulation.* 2011;123:1594-1600).

Assuming a 100% effective intervention, sickle trait screening was estimated to potentially avert approximately seven sudden deaths in NCAA athletes over a 10-year period, almost all in football, basketball and track (one per 144,000 athletes) at a cost of $1.4 million to $2.8 million per event (Tarini BA, et al. *Health Serv Res*. 2012;47:446-461). These authors also point out that even more lives potentially could be saved with cardiovascular screening, which is more complicated and expensive.

Two divergent viewpoints remain concerning the wisdom of screening college athletes for sickle trait. Those in favor cite the increased rate of unexplained death in individuals with sickle trait and suggest that targeted intervention would be effective. Those who oppose screening recommend universal modification of training practices for all athletes. The Army eliminated the increased rate of death in persons with sickle trait with universal modifications to basic training and no longer screens recruits (Tarini BA, et al. *Health Serv Res*. 2012;47:446-461).

Studies consistently indicate that sickle trait is identified in only a small minority of episodes of unexplained death. Multiple other conditions, some of which are much more common such as obesity, concurrent illness, diabetes and cardiovascular disease, also are frequent risk factors.

The American Society of Hematology's *Statement on Screening for Sickle Cell Trait and Athletic Participation,* endorsed by the American

Society for Pediatric Hematology/Oncology, recommended the use of universal modifications instead of sickle trait screening (http://hematology.org/advocacy/policy-statements/7704.aspx). It cites concerns that mandatory trait screening would lead to selection discrimination against the athlete with sickle trait. However, the NCAA policy states that athletes with sickle trait should be allowed to participate in athletics. Few data exist that support or refute the concerns of discrimination related to sickle trait testing.

Screening for sickle cell trait

Newborn screening for sickle hemoglobin began in the 1980s in many states and has been performed routinely in all 50 states since 2006; hence, most adolescents already have been screened for sickle cell trait. The pediatrician should review these results with adolescents during health care visits and advise the patient and family about the genetic risk as well as sensible precautions for athletic participation. Universal newborn screening has educated physicians that sickle cell trait may occur in individuals from India, Central/ South America, the Middle East, Eastern Europe as well as those from Africa.

The most commonly advocated technique for trait screening is the solubility index (Sickledex). While this technique has a reported sensitivity to identification of sickle hemoglobin of 98.9%, it does not distinguish between sickle trait and forms of disease such as Hb SS, Hb SC, Hb Sb thalassemia. These conditions have potential for significant acute and chronic manifestations. Therefore, a positive solubility test should be followed with an Hb electrophoresis to exclude more significant hemoglobinopathies. The financial estimates per life saved do not include the significantly increased cost of the appropriate follow-up testing.

Counseling patients

Sudden death in sickle trait appears to occur during early training conditioning exercises and not necessarily during competition. Deaths are associated with high levels of exertion without prior training and under adverse environmental conditions such as excess heat, dehydration or altitude. While exactly how much added risk sickle trait causes for exertional rhabdomyolysis is unclear, pediatricians will be asked to counsel adolescents with sickle cell trait about the risks of their status. This requires that the pediatrician have:

- access to sickle testing results — either newborn screening results or a new testing protocol as above;
- knowledge of the various sickle syndromes and their distinction from Hb AS (sickle trait); and
- an understanding of exercise modifications that decrease the risk of environmental stress-related illness.

With this in mind, pediatricians should follow recommendations of the AAP policy on *Climatic Heat Stress and Exercising Children and Adolescents* for all high-risk individuals if not for all athletes. Additionally, all coaches, athletic trainers and people overseeing exercise programs for children and adolescents should be aware of the early signs of heat-related illness and responses to identification of these conditions in those under their care.

Athletes and their parents should be encouraged to know their individual status with regard to sickle cell trait whenever testing results are available and what concerns or precautions may be necessary to promote a healthy, active lifestyle.


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