Sickle Cell & The Athlete

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No Disclosures

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Michigan v Villanova – Elite Eight 2022

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Objectives

- Review etiology of Sickle Cell Disease and Trait
- Identify differences between SCD and SCT
- Discuss risks faced by a SCT athlete
- Understand activity modifications for the SCT athlete

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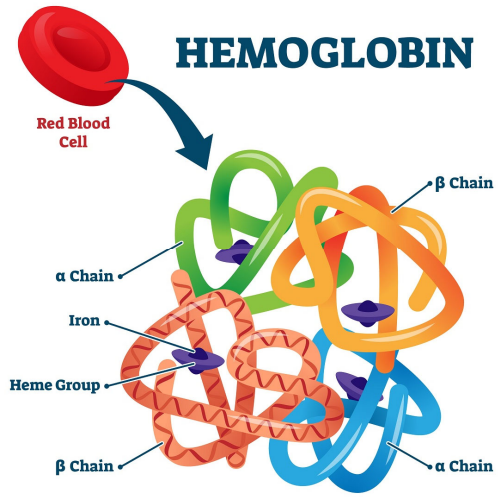
Etiology

- Hemoglobin
  - Two alpha chains
  - Two beta chains

  - A – normal
  - S – abnormal
    - valine substitution for glutamic acid



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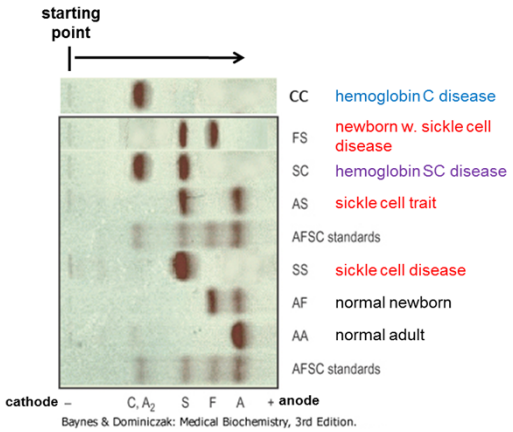
Etiology

Laboratory Evaluation

- Sickle Prep Test
  - Blood smear
- Sickledex
  - Hgb solubility
- Hgb electrophoresis
  - **Gold Standard**
  - Can give % of types of Hgb
  - Can **distinguish** an individual **SCD vs SCT**



Baynes & Dominiczak: Medical Biochemistry, 3rd Edition.

A: normal hemoglobin β chain (HbA)
F: normal hemoglobin γ chain (HbF, fetal)
S: sickle cell hemoglobin β chain (HbS)
C: hemoglobin C β chain (HbC)
A_2: normal hemoglobin δ chain (HbA_2)

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Etiology

Sickle Cell **Disease**

- Two normal alpha chains
- Two Hgb S beta chains

- Hgb S **sickles** with deoxygenation
- Susceptible to anemia and painful crisis
- Normally **"non-athletes"**

Sickle Cell **Trait**

- Two normal alpha chains
- One normal Hgb A beta chain
- One abnormal Hgb S beta chain (30—44% concentration)

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Sickle Cell **Trait**

- Epidemiology
  - 1.6% US population
  - Around 8% of AA population
- Normally ASYMPTOMATIC
  - **Unless Stressful environment**
    - Extreme heat
    - High Altitude

- Screening
  - All states require neonatal screening
  - **Required at the NCAA Div 1 & 2 level**
    - ACSM & NCAA joint statement – March 2012
    - Possible to opt out

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The Sickle Cell Trait Athlete

Overexertion is associated with exertional collapse and sudden death

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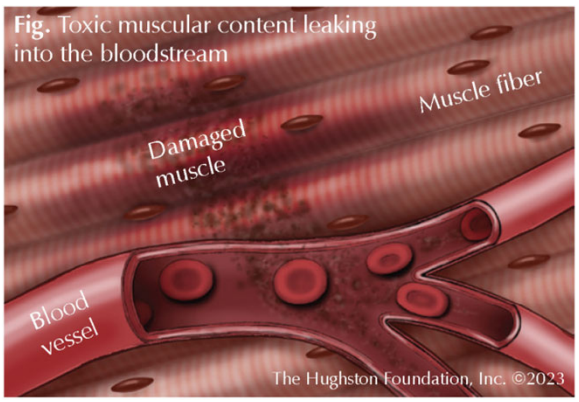
Sickle Cell Trait - Complications

- Rhabdomyolysis
- Exertional Heat Stroke
- Exertional death associated with SCT



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Sickle Cell Trait

- **Rhabdomyolysis**
  - Exercise induces deoxygenation and sickling, *there is spectrum of intensity and response*
    - Uncompensated hypoxia -> sickling
    - Maximal exertion -> mild sickling
    - Increased altitude -> sickling
  - Sickling leads to **vaso-occlusion and muscle pain**
- Can lead to **necrosis and extravasation of contents**
  - Increased CK
  - Myoglobinuria
  - Myalgia
  - Renal damage/failure
- Muscles are **not usually tense or hard** (like cramping)
- Quicker?

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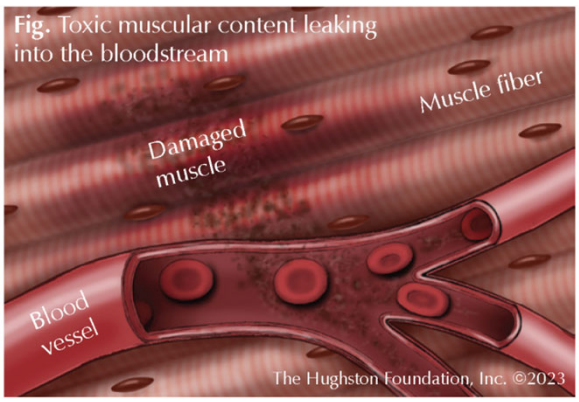
Sickle Cell Trait

Rhabdomyolysis

- Treatment
  - Recognition
  - Vital signs
  - Supplemental oxygen
  - Cool athlete
- Implement **emergency response** if necessary
  - IV fluids



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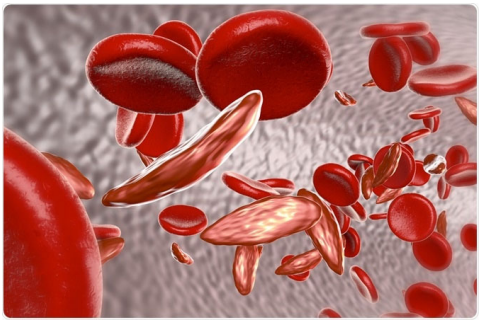
Sickle Cell Trait

Exertional Heat Stroke

- High core temp increased sickling

Exertional Death assc w/ SCT

- Absolute risk is small
- However, risk of sudden cardiac death is 40% higher than non-SCT



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The Sickle Cell Trait Athlete

- Increased risks
  - Splenic infarction
    - LUQ pain, nausea vomiting
  - Hematuria
    - Sickling in renal medulla
    - Possible papillary necrosis
  - Hyposthenuria
    - Inability to concentrate urine
      - Cumulative effects of papillary necrosis
      - Can contribute to **dehydration**
      - Dehydration -> sickling

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Exercise Guidelines

- Goal: Prevent overexertion and dehydration while gradually acclimatizing athletes
  - Encourage preseason training and conditioning
  - Gradual progression of exercise
    - Allow for longer periods of recovery between reps
- Reported higher incidence of EAC on Day 1 of conditioning in the summer
- Training modification
  - Acclimatize to temp and altitude
  - Avoid dehydration or overexertion
  - Gradual progression in workout intensity
  - Longer recovery periods in workout/reps
  - Don't work out when sick (esp fever)

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COMPETITIVE SPORTS AND PAIN MANAGEMENT

Sickle Cell Trait in Sports

E. Randy Eichner
Professor Emeritus of Medicine, University of Oklahoma Health Sciences Center, Oklahoma City, OK

EICHNER, E.R. Sickle cell trait in sports. *Curr. Sports Med. Rep.*, Vol. 9, No. 6, pp. 347–351, 2010. *Sickle cell trait (SCT) can pose a grave risk for some athletes. In the past decade in NCAA Division I football, no deaths have occurred from the play or practice of the game, but 16 deaths have occurred from conditioning for the game, and 10 (63%) of these deaths are tied to SCT, an excess of up to 21-fold. Research shows how and why, during intense exercise bouts, sickle cells can accumulate and "logjam" blood vessels, causing explosive rhabdomyolysis that can kill. Sickling can begin in 2 to 5 min of all-out exertion and can reach grave levels soon thereafter if the athlete struggles on or is urged on by coaches despite warning signs. Sickling collapse is an intensity syndrome that differs from other common causes of collapse. Tailored precautions can prevent sickling collapse and enable athletes with SCT to thrive. Irrationally intense conditioning for a game puts the lives of healthy athletes with SCT at risk.*

Original article

Sickle cell trait associated with a RR of death of 37 times in national collegiate athletic association football athletes: a database with 2 million athlete-years as the denominator

Kimberly G Harmon,[1] Jonathan A Drezner,[1] David Klossner,[2] Irfan M Asif[3]

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Which of the following statements regarding sickle cell trait athletes is true? *(AMSSM Sports Medicine CAQ Study Guide, 2nd Edition)*

- Sickle cell trait, in contrast to sickle cell disease, has little to no mortality in athletes
- Any cramping, struggling, or collapse in a sickle cell trait athlete must be considered sickling, a medical emergency, until proven otherwise
- The symptoms of exertional sickling and heat illness (heat stroke or heat cramping) are not distinguishable
- Acclimatization to intense training, increased hydration, and increased rest afford no protection to sickling in athletes

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During a preparticipation examination of an NCAA Division I college soccer you discover he is positive for sickle cell trait. Which of the following is statement concerning the prevention of complications in this athlete? *(AMSSM Sports Medicine CAQ Study Guide, 2nd Edition)*

- No special recommendations are needed as the patient has sickle cell trait not sickle cell disease
- The athlete should push through muscle pain and undue fatigue while conditioning, as these are likely signs of poor endurance from lack of a proper off-season training program
- The athlete should modify activity only after altitude of competition exceeds 5,000 feet
- The athlete should avoid all-out exertion of longer than two minutes without a rest interval

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You are starting a new position in the state of Colorado as a high school team physician. Towards the end of the first football practice of the year, during conditioning drills, an African-American football player collapses to the turf. He complains of severe pain in his bilateral quads and tells you that he is cramping.
As the new team physician, you don't know the players yet. You ask the trainer about his medical history, and his response is that this athlete is a recent transfer student from Alabama, and he doesn't know him well. The athlete doesn't know any of his past medical history, other than to say that he's pretty healthy and doesn't know any of his family history. On exam, his muscles are soft, and there is no spasm. The cramps are spreading. He denies any new medications or drug use. He is lucid, alert, and oriented. What should your working diagnosis be, and what is your next step? *(AMSSM Sports Medicine CAQ Study Guide, 4th Edition)*

- Cramping from dehydration-start an IV or give him oral fluids
- Sickle cell trait-start high flow oxygen and call 911
- Heat stroke-get him in a tub of cold water and call 911
- Not in shape give him some water and let him sit on the sideline until he recovers, then let him go back to practice

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What about Sickle Cell **Disease** Athlete?

- We want to promote healthy exercise in all individuals.
- Individual assessment, case-by-case
- **Increased risk with strenuous activities, contact and collision sports.**

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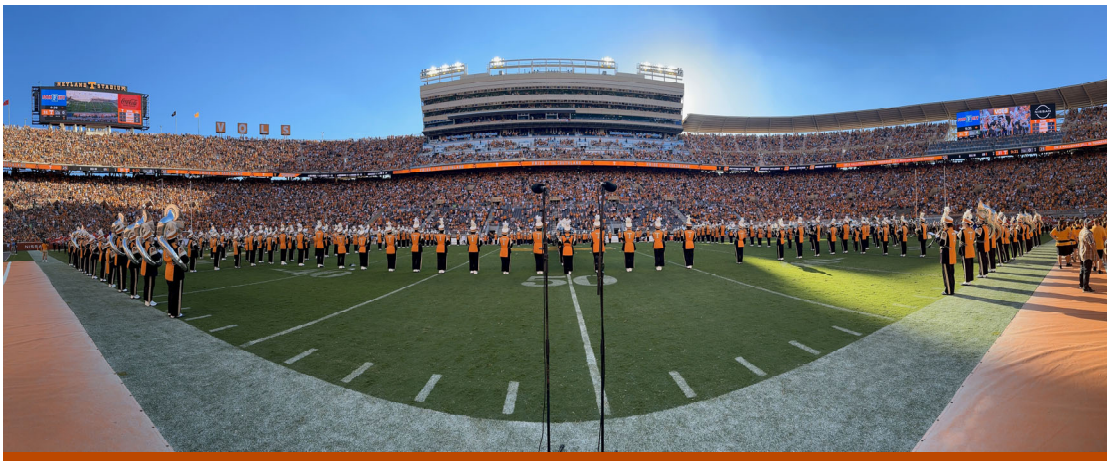

UTSA v Tennessee at Neyland Stadium. 9/23/23

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