

Protect Life: The Athlete with Sickle Cell Trait

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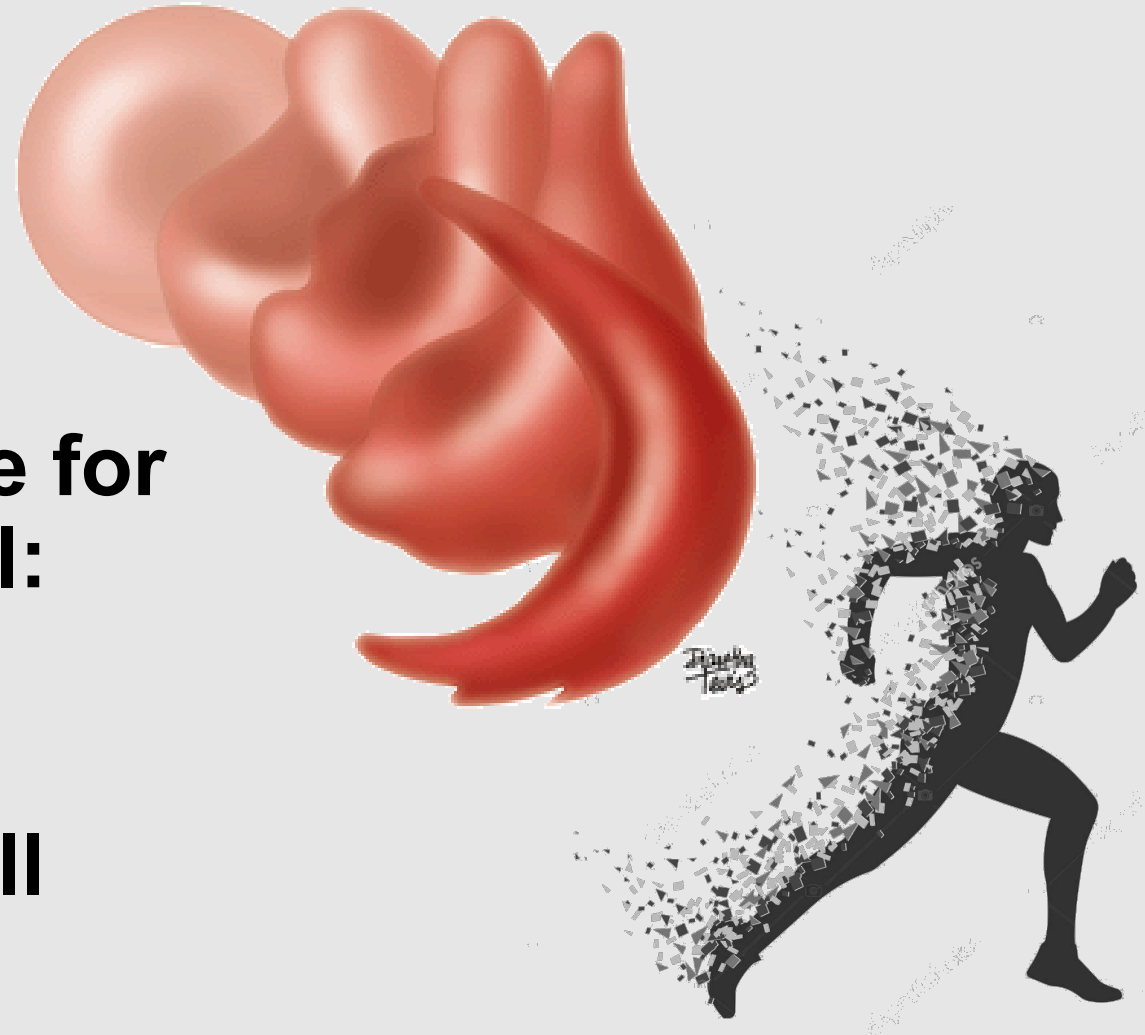
Disclosures

I have no actual or potential conflict of interest in relation to this program/presentation.



What is Sickle Cell Trait?

- **Not a disease state**
 - Normal life span
 - No anemia
- **A condition of inheritance**
- **Inheriting one abnormal gene for hemoglobin (S), each red cell:**
 - ~ 40% hemoglobin S
 - ~ 60% hemoglobin A
- **Genetic type AS vs Sickle Cell Anemia SS**



Eichner's Quintus

1. Asymptomatic Sickle Cell Trait
2. Ischemic events
3. Ischemic events with visceral or muscular infarcts
4. Severe rhabdomyolysis with residuals (kidney disease, splenic disease, MI)
5. Fulminant rhabdomyolysis with exertional sickling (ES) death



ES Deaths



2 MMA/Kickboxers



7 Recruits



1 High School



1 PE Class

**3 Male
2 Female**



2 Frat Pledges



**26 College
16 High School
2 Jr High
1 Youth**



**59 Police
Custody**



**6 Boxers
2 Fistfight**



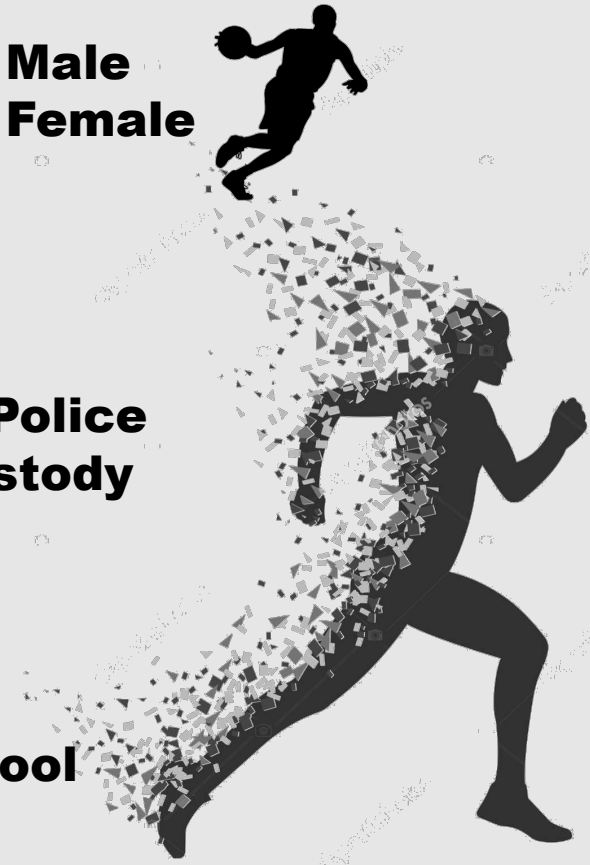
11 Firefighters



**14 Air Force
32 Army
10 Navy
1 USCG
1 Marine**



**1 High School
1 College**





- **Consensus Statement: Sickle Cell Trait and the Athlete [2007]**
- **Position Statement: Preventing Sudden Death in Sport [2012]**
- **Preventing Sudden Death in Collegiate Conditioning Sessions: Best Practice Recommendations [2012]**
- **Preventing Sudden Death in Secondary School Athletics Programs: Best Practice Recommendations [2013]**



- **Athletes should know their SCT status**
- **Strength Coaches, sport coaches and medical staff should be aware of the SCT status of each athlete**
- **The supervising staff should know prevention, recognition and treatment strategies for exertional collapse in athletes with SCT**



Casa DJ, Anderson SA, Baker L, et al. The inter-association task force for preventing sudden death in collegiate conditioning sessions: best practices recommendations. J Athl Train. 2012 Jul-Aug;47(4):477-80.

Casa DJ, Almquist J, Anderson SA, et al. 2013. The inter-association task force for preventing sudden death in secondary school athletics programs: best-practices recommendations. J Athl Train 48:546-553.

Prevention Strategies

Knowledge Beats Ignorance & Prevention Beats Treatment

1. Know SCT status
2. Targeted Education: the athlete and the principal staff
3. Implement Tailored Precautions
4. Detect ES warning signs & symptoms *very early*
5. *Stop exercising immediately!*
6. Assess

There is no evidence that any player has ever died an ES death as SCT status is known, athlete and staff are educated, and precautions are heeded



Precaution Strategies

“Don’t let the finish line kill that warrior!” Cliff Melton

Exclude from Day-1 conditioning tests

Slow, paced progressions of training

Allowing longer periods of rest and recovery between repetitions

Heat stress, dehydration, asthma, illness, and newness to altitude create additional risk

Stop activity with onset of symptoms

Set a tone that encourages consideration for the athlete with sickle cell trait

Act fast in collapse



Common ES Settings

- **Windsprints**
- **Timed miles**
- **Uphill repeats**
- **Ramp running**
- **Mat drills**
- **Weight training**
- **Resistance drills**
- **Police Cadet – ‘defensive tactics drill’, timed distance run**
- **Firefighter – ‘tower drill’, timed distance run**
- **Military – ‘fitness’ testing**



Recognition Strategies

Sickling Collapse: Unique?

There Are Telltale Clues...

Brief, but sustained, “heroic” exercise

‘Fatigue’

Legs like Jell-O

“My chest hurts”

“I can’t catch my breath”

Ischemic pain / ‘cramp’

Low back pain, ‘cramp’, spasm

Leg pain, ‘cramp’, spasm

Hyperventilating from lactic acidosis

DO NOT ASSUME A PRODROME!

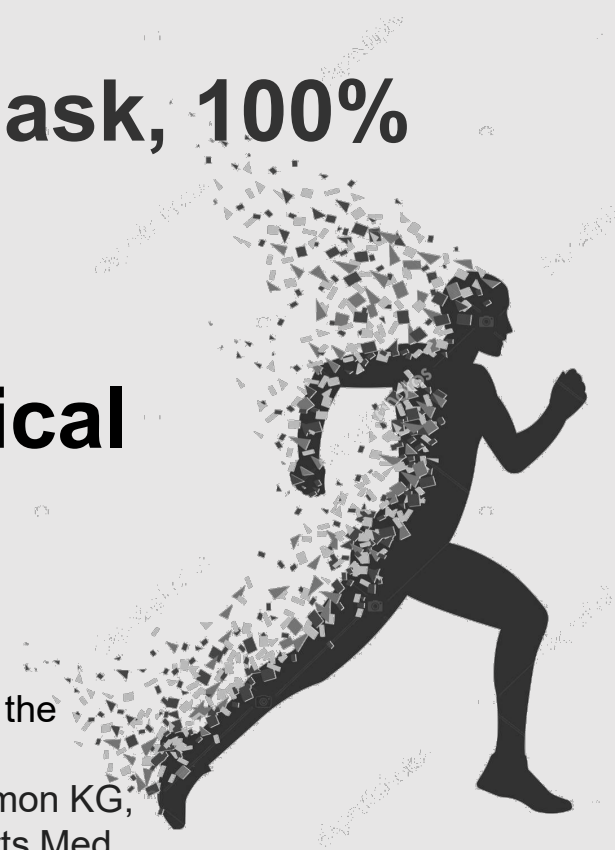


Treatment Strategies

- **Proactively prepare by having an EAP and appropriate emergency equipment**
- **Rapid recognition**
- **O₂**
 - **High flow 15 lpm with non-rebreather mask, 100% O₂**
- **AED**

In the event of ES collapse, treat it as a medical emergency

- **Check vital signs**



Interassociation Task Force on Sickle Cell Trait and the Athlete. (2007). Consensus Statement: Sickle Cell Trait and the Athlete. National Athletic Trainers Association

O'Connor FG, Franzos MA, Nye NS, Nelson DA, Shell D, Voss JD, Anderson SA, Coleman NJ, Thompson AA, Harmon KG, Deuster PA. Summit on Exercise Collapse Associated with Sickle Cell Trait: Finding the "Way Ahead". Curr Sports Med Rep. 2021 Jan 1;20(1):47-56.

Racial Bias In Pulse Oximetry

High flow 15 lpm with non-rebreather mask, 100% O₂

▪ ***Even if normal pulse ox***

Pulse ox data from case reports:

- 81%, 77% (for 2) and even as low as 62%

Pulse ox can "overestimate" arterial O₂ sat during hypoxemia in Blacks (vs Whites)

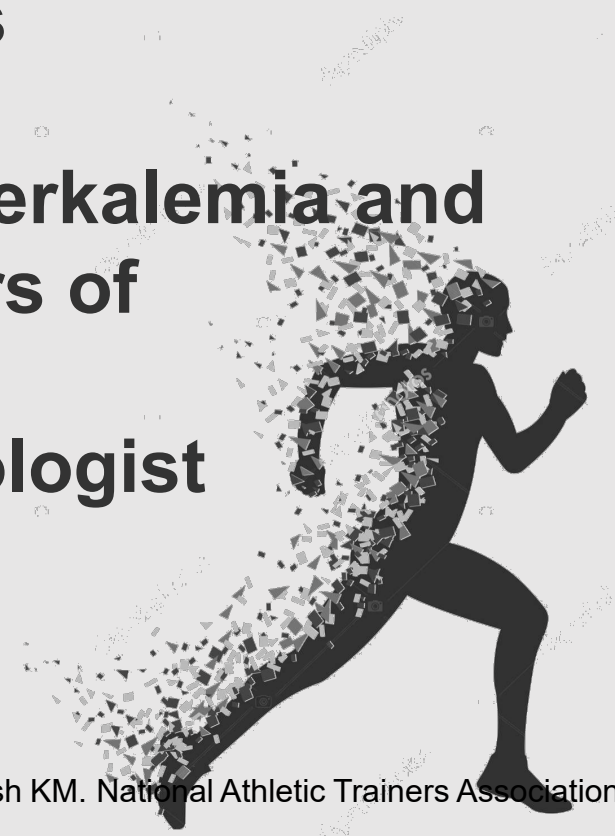
In other words, the pulse ox reading for O₂ sat in Blacks can be 4-5 points higher than the arterial blood O₂ sat



Emergency Room Strategies

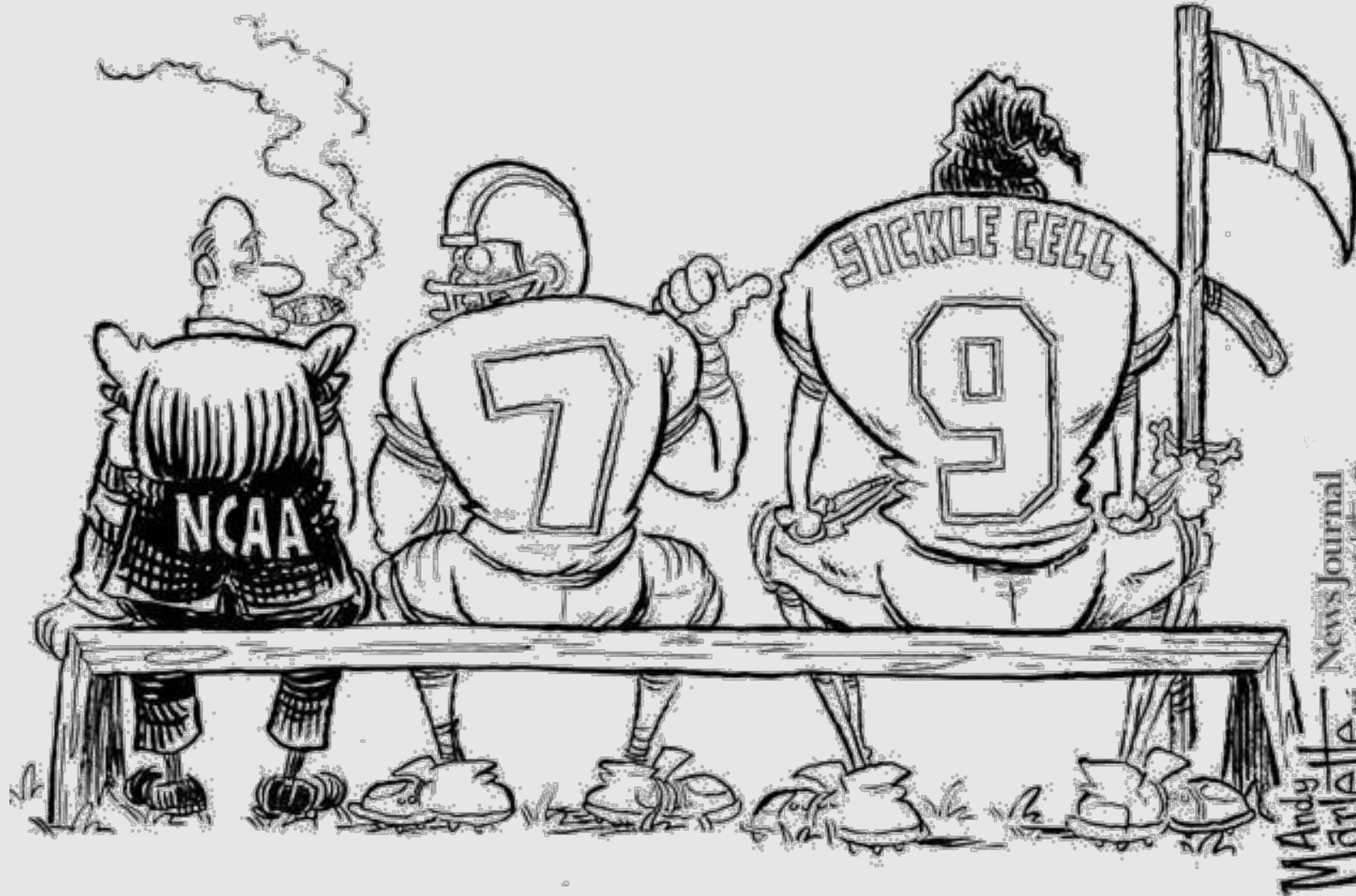
- Inform treating physicians of SCT status so that they are prepared to manage explosive rhabdomyolysis and associated metabolic complications
- Aggressive fluid and electrolyte management
- Blood gas monitoring to rule out metabolic acidosis
- Cardiac monitoring – Cardiologist
 - Patients with severe ES may rapidly develop hyperkalemia and lethal cardiac arrhythmias within minutes to hours of presentation
- Possibly dialysis to control myoglobinuria – Nephrologist
- Anticipate fasciotomy – Orthopedic Surgeon

Generalizing **broadly**...from our data...the chance of mortality is ~70% in full-out ES collapse



There Is No Contraindication To Participation In Sport For The Athlete With SCT!

"SERIOUSLY?! YOU'RE GRANTING THIS GUY ELIGIBILITY?!?!"



ES Case Study: Eric Goll, Chadron State College

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Erick Goll

- **20 y/o black male**
- **330 pound Defensive Lineman**
- **Florida native**
- **2014 – Year one football player, Chadron State College (CSC) Nebraska**
- **2015 – Year two football player, Florida A&M University (A&M)**
- **2016 – Year three football player, CSC**



Aug 2016 – Year 3 – CSC

Aug 10

- Eric signs a waiver declining PPE test for SCT
- Checked “yes” to PPE sickle cell question
- No documented medical clearance for athletic participation

Aug 11

- Day-1 Conditioning Test, 300-yard shuttles
- Became fatigued and ‘cramped’ after two 300-yard runs
- Given water, cold towel, and stretched by ATs
- Subsequently transported to cold tub
- 2 minutes after CWI became unresponsive
- EMS already contacted re: a different player ‘cramping and shivering’
- Transported to hospital
- Pronounced dead at 12:30pm; ~1 hour after LOC



Past Medical History

2014 – CSC

- PPE – no test for SCT & no waiver for testing
- Day-1 conditioning test
- CSC Head AT: “same shit” [fatigue & ‘cramping’] as 2016; thought he just overheated
- Treated with cold tub, Gatorade, water; ate and was fine

2015 – A&M

- 7/20/15 – tests positive for SCT
- Multiple EKGs, a cardiac stress test and echocardiogram
- Cleared for athletic participation



“Learn from the mistakes of others. You can't live long enough to make them all yourself.”

Eleanor Roosevelt

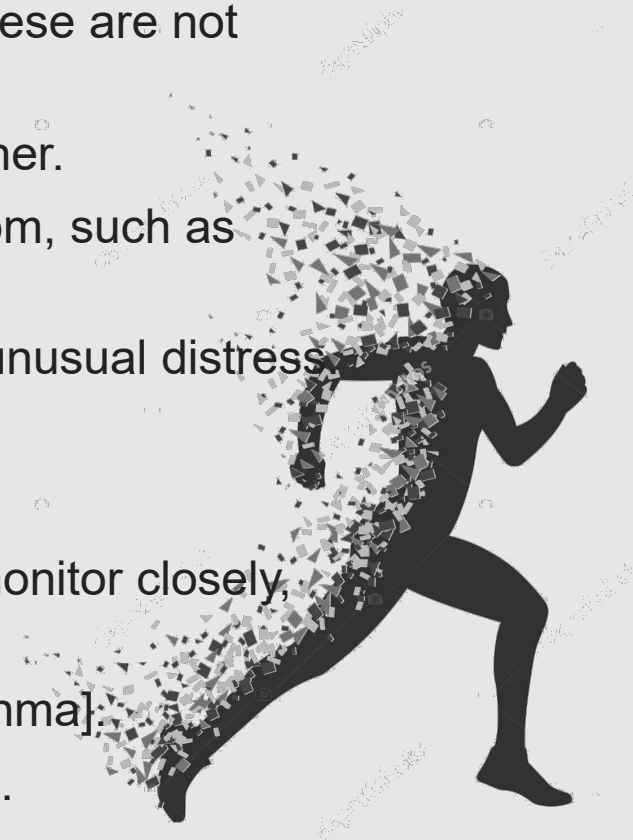
Take-home Lessons:

- **Confirm SCT Status in the PPE**
 - **NCAA waiver eliminated effective Aug 2022**
- **Educate the Athlete and all ‘Stakeholders’**
 - **Athlete**
 - **AT, S&C coaches, coaches, team physicians, parents**
- **Implement Precautions**
- **Know characteristics of collapse**
- **Treat ES collapse as a medical emergency**
- **Abolish ‘conditioning tests’ lacking specificity and sensitivity**



Precautions for the Athlete with SCT

1. Ideally, athletes with sickle cell trait should set their own pace.
2. Engage in a slow and gradual preseason conditioning and lifting regimen.
3. Build up intensity slowly while training, e.g., paced progressions.
4. Use adequate rest and recovery between repetitions, especially during serial sprints and intense station or mat drills.
5. Not to participate in “preseason conditioning tests.”
6. Be excused from performance tests such as timed serial sprints or miles, especially if these are not normal sports activities.
7. Not be urged to perform all-out exertion of any kind beyond 2-3 minutes without a breather.
8. Stop activity immediately upon struggling, i.e., at the earliest onset of any undue symptom, such as cramping, pain, weakness, breathlessness, or fatigue.
9. Report any symptoms to athletic trainer and coach. Seek prompt medical care for any unusual distress.
10. Stay well hydrated at all times, especially in hot and/or humid conditions.
11. Curb activity in very hot or humid conditions.
12. Curb activity when new at altitude, even a “jump” of only 2,000 ft. Cut training effort, monitor closely, have ready access to supplemental oxygen.
13. Control asthma to cut risk of exertional sickling [... has / does not have a history of asthma].
14. Decrease activity sharply during any illness, especially with fever, vomiting, or diarrhea.
15. Decrease activity after nights of poor sleep.



Characteristics of Non-Traumatic Collapse

NON-TRAUMATIC COLLAPSE				
	Exertional Sickling	Cardiac	Exertional Heat Stroke	Asthma
Onset	Can occur early in workout	No warning	Usually occurs late in workout	Usually known asthma
	Slumps to ground	Unprotected fall	Can be in coma	Prior episodes, poor control
	Slow evolution to a conscious collapse	Unconscious and unresponsive		Usually occurs after sprinting
Signs & Symptoms	Responsive, initially	Unresponsive	Fuzzy thinking	Breathless, may/not wheeze
	Weakness > pain; low back and/or legs	Gasping, gurgling, snorting, or moaning	Bizarre behavior	Gasping, panicky, on hands/knees
	No palpable muscle cramp	Limp or seizing	Incoherent	Auscultate: moving little air
	Core Temperature < 103° F	Core Temperature irrelevant	Core Temperature > 106° F	Core Temperature irrelevant
Management	High-flow O ₂ , 15 lpm non-rebreather	CPR, AED	Cold water immersion	Epi-pen, O ₂



You Haven't Seen ES...Has ES Seen You!?

CSC 2015

- **Player with SCT**
- **'overexerted himself'**
- **Symptoms greater than Eric**
- **Cold tub, then shower**
- **Coach called Head AT: kid "going nuts"**
- **Transported to hospital**
- **Dx: Rhabdo; hospitalized 8 days with 21 L IVF**
- **Reportedly never returned to athletic participation at CSC**



Confirming SCT Status in the PPE

“The preparticipation physical examination provides a key opportunity for counseling a SCT positive athlete with specific education for risk of exertional sickling prior to commencement of strenuous athletic participation.”

Taylor C, Kavanagh P, Zuckerman B. Sickle cell trait--neglected opportunities in the era of genomic medicine. *JAMA*. 2014;311(15):1495-1496.

“Due to high proportion of student-athletes who are unaware of their SCT status, institutions should facilitate SCT screening with confirmatory testing for all student-athletes to prevent missed identification of those with SCT.”

Hirschhorn RM, et al. The Prevalence of Sickle Cell Trait in Division I University Athletics Programs.: *Physician and Sportsmedicine* 2020 Mar 12;1-7

“The SCT screening bylaw adopted by the NCAA in 2010, with associated precautions, has successfully prevented ES with SCT fatalities in NCAA Division I football players. Similar SCT guidelines and enforced compliance should be mandated in all sports at all levels.”

Boden BP, Fine KM, Breit I, Lentz W, Anderson SA. Nontraumatic exertional fatalities in football players, part 1: epidemiology and effectiveness of national collegiate athletic association bylaws. *Orthop J Sports Med*. 2020 Aug 19;8(8)

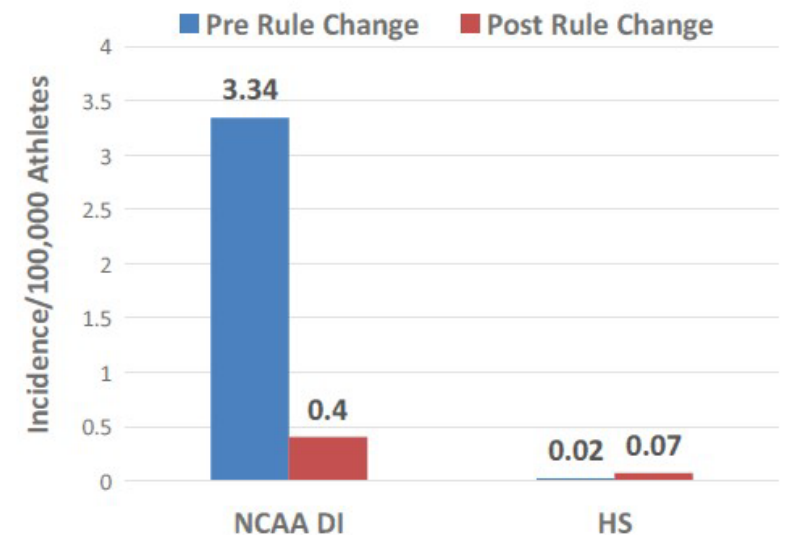


Figure 5. The incidence of exertional sickling with sickle cell trait (SCT) fatalities per 100,000 athletes before and after the National Collegiate Athletic Association (NCAA) 2010 SCT screening bylaw. DI, Division I; HS, high school.

Resource Videos

‘Melton video’: [Sickle Cell Trait Implications on Exercise and Military Service – YouTube](#)

Performance Training Strategies to Prevent Catastrophic Injuries and Death: [Performance Training Strategies to Prevent Catastrophic Injuries and Death \(CATS\) - YouTube](#)





The End!

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