SICKLE CELL TRAIT
CONCEPTS AND CONTROVERSIES

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Sickle Cell Trait

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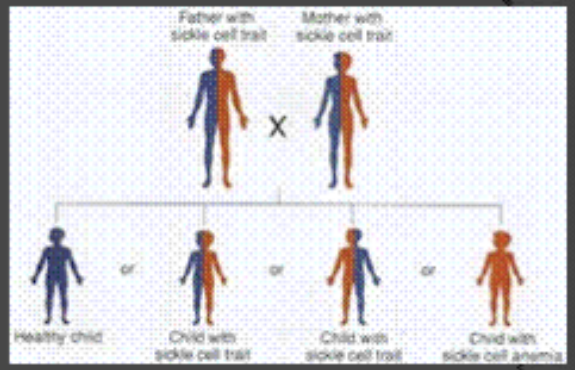
Sickle Cell Trait

- A hereditary condition in which an individual inherits one normal gene for hemoglobin (A) and one abnormal gene for hemoglobin (S)
- Differs from Sickle Cell Disease in which the individual inherits two abnormal genes for hemoglobin (SS)



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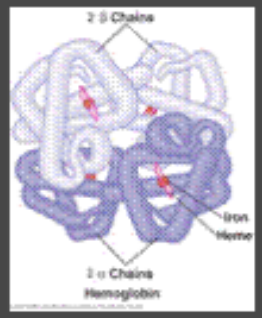
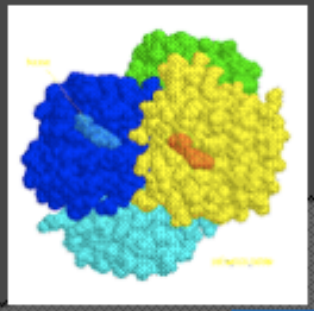
Hemoglobin

- Hemoglobin is a protein in red blood cells that is responsible for binding oxygen and carrying it throughout the body to be used somewhere else and then carrying carbon dioxide away from cells to be released in the lungs.
- Hemoglobin is made up of 4 chains of amino acids: 2 alpha chains of 141 aa each and 2 beta chains of 146 aa each with an iron molecule in the center





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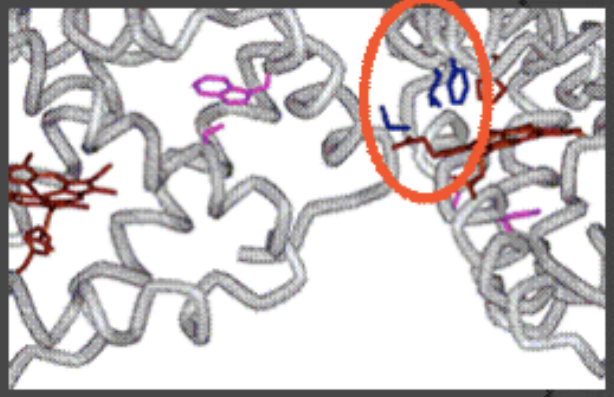
Hemoglobin S

- A gene mutation that changes one amino acid (valine for glutamic acid at the 6th position) in the beta globulin chain causes the molecule to have a slightly different shape that allows the molecule to become "sticky". The molecules then stick together and change the shape of the cell especially when there is little oxygen



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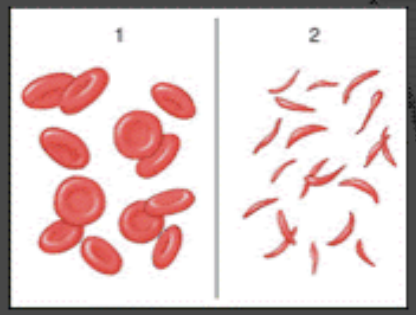
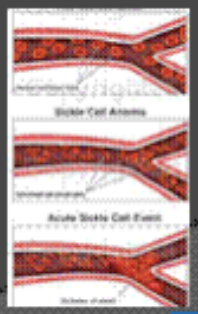
Sickle Cell Disease

- When oxygen level is low, the shape of the cell changes causing it to become stiffer
- This change in shape causes the cells to get stuck in small blood vessels and prevents blood flow





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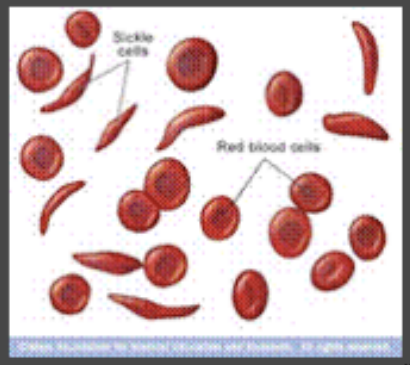
Sickle Cell Trait

- Affects 3 million Americans
- Occurs in 1 in 12 African Americans (8-10%)
- Occurs in 1 in 2,000-10,000 Caucasians
- Also present in:
  - Mediterranean
  - Middle Eastern
  - Indian
  - Caribbean
  - Central and South America



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Sickle Cell Trait

- First recognized in the military in 1970 with the death of 4 recruits at Fort Bliss,TX (alt: 4,060ft).
- They concluded that an individual with sickle cell trait was 30 times more likely to die during basic training.
- First athlete death reported in 1974
- The cause of death in 16 college football players
- Has been linked to the death of 11 athletes in the past 8 years (7 college football, 2 high school, 2 12 year olds)

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Sickle Cell Trait

- The cause of death is from exertional rhabdomyolysis, a condition of muscle break-down that leads to kidney failure and other catastrophic events and exercise-associated sudden death
- The risk of exertional rhabdomyolysis is 200 times greater for an individual with Sickle Cell Trait

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Sickle Cell Trait

- The risk of problems associated with Sickle Cell trait are increased with:
  - Extreme Heat
  - Dehydration
  - Altitude
  - Asthma
  - Drugs

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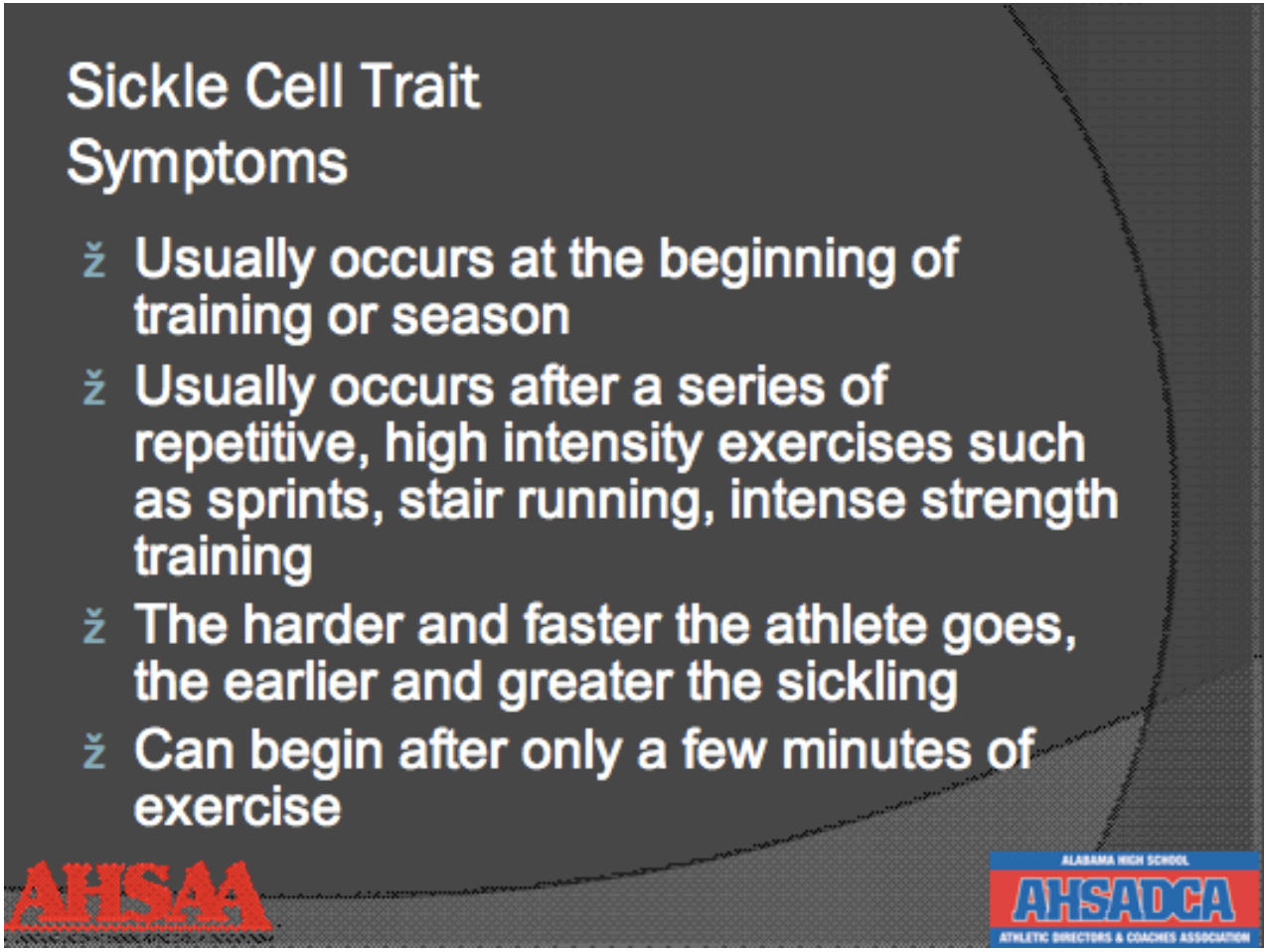
Sickle Cell Trait
Symptoms
Usually occurs at the beginning of training or season
Usually occurs after a series of repetitive, high intensity exercises such as sprints, stair running, intense strength training
The harder and faster the athlete goes, the earlier and greater the sickling
Can begin after only a few minutes of exercise
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Sickle Cell Trait Symptoms

- Differs from Heat Exhaustion:
  - Occurs in the beginning of activity
  - Core temperature not elevated
  - Not associated with muscle cramps, but do have muscle pain
  - Athletes don't "lock-up", but slump to ground
  - Respond quicker to proper treatment than heat crampers

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Precautions and Treatment

- No athlete is disqualified
- Build up slowly with paced progressions
- Allow longer periods of rest and recovery between repetitions
- Encourage pre-season conditioning
- Athletes with SST should not do performance testing (timed runs/sprints)

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Precautions

- Stop activity if any symptoms:
  - Muscle pain
  - Weakness
  - Muscle swelling
  - Inability to “catch breath”
  - Severe fatigue
  - Collapse

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Precautions

- Adjust workouts if extremely hot (see the NATA's Preseason Heat-acclimatization Guidelines for Secondary Schools)
- Hydrate
- Control asthma
- Hold sick athlete out of workout
- Modify activity at altitude
- Educate athlete, coaches, and other personnel to report symptoms early

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Treatment

- Recognize the problem
- Protect the athlete- from environment, coaches, other athletes
- Check vital signs
- Administer oxygen at high flow with a non-rebreather mask
- If signs of collapse/obtunding or worsening condition, activate emergency plan
- I.V. fluids

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Controversies

- Mandatory testing of all athletes:
  - Cost- $30/test
  - Discriminatory
  - May miss athletes with trait that are not of African decent
  - 64% of NCAA Division I-A schools test
  - NFL Scouting Combine screens
  - Potential legal ramifications

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Testing at Alabama

- 379 = athletes on scholarship
- 604 = total number of athletes
- 147 = total number of African decent athletes
- 36 = total number of new African decent athletes
- $4,500 = start up cost of targeted testing
- $1,080 = yearly cost of targeted testing

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Recommendations

- Treat ALL athletes as if they have Sickle Cell Trait
- Identify athletes with known SST
- Get records from State Health Departments as ***all*** states now require screening of ***all*** newborns. First started in 1975. In 1999, 4 states still did not test (Utah, Montana and Dakotas) and 5 others only did targeted testing (MA,NH,WV,ID,HI)
- Incorporate results of Newborn screening on the Pre-Participation Exam

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Recommendations

- All newborn screening is done by isoelectric focusing (IEF) from dried blood samples
- Positives confirmed by hemoglobin electrophoresis with citrate agar

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