

Students with Sickle Cell Disease

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Students with Sickle Cell Disease

Transition from
Hospital
To home
To School

Definition:

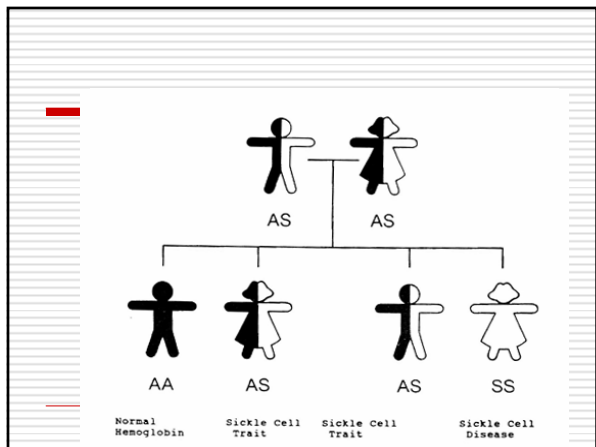
- The sickle cell diseases are a group of hereditary hemoglobin disorders characterized by red cells that undergo a sickle shape transformation when they are deoxygenated.
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Incidence:

- ❑ The sickle gene is most prevalent in Africans, Arabs, Egyptians, Turks, Greeks, Italians, Iranians, Asiatic Indians.
- ❑ Rarely occurs in caucasian population

African-Americans:

- ❑ 1 out of 12 is a carrier of sickle cell
- ❑ 1 out of 600 have the disease



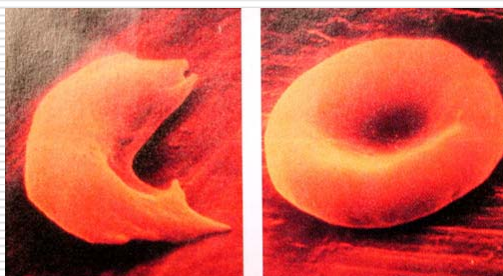
Types of Sickle Cell Disease

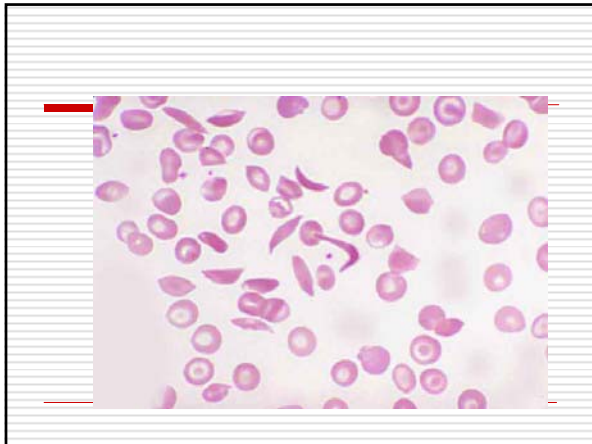
- ❑ Sickle cell Hemoglobin SS- considered more severe form
- ❑ Hemoglobin SC disease
- ❑ Sickle beta-thalassemia
- ❑ Others: Sickle cell trait, etc.

Differences in Red Blood Cells

- | | |
|----------------------------------|--|
| ◆ Normal RBC: | ◆ RBC containing Hgb S: |
| ◆ Pliable, biconcave disc | ◆ Distorted shape: crescent/sickle shape |
| ◆ Life span: 120days | ◆ Life span: 10-20 days |

Sickle Cell Normal RBC





Factors That Influence Cells to Sickle:

- Fever
- Infection
- Dehydration
- Sometimes no apparent cause

All Problems Related to Sickle Cell Anemia are related to:

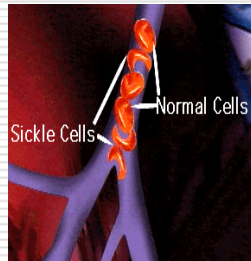
- Anemia (hemolytic)
- Vascular Occlusion
 1. Pain
 2. Organ Dysfunction
 3. Organ Failure
 4. Susceptibility to infection
- Psychosocial problems
- Growth failure

Results of "Sickling":

- ❑ 1. Anemia
- ❑ 2. Vaso-occlusive complications

Vascular Occlusion

- ❑ Sickle cells are sticky and tend to stick together. Their sickle shape and rigid structure makes it difficult to pass through vessels.





Pain Crisis

- Can occur anywhere in the body.
- Common sites: legs, back, abdomen.

How to Handle Pain Crisis at School:

- If pain is mild: Tylenol, Motrin, rest, and drinking extra fluids.
- If pain is mild: Coping strategies such as distraction, calming statements.
- Do not try to talk the child out of his pain.
- If pain is severe will need professional intervention.
- Ask the child what "works" for his pain.

Anemia

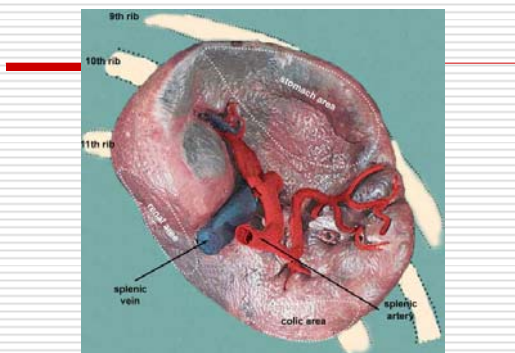
- Most children will have a hemoglobin lower than normal all the time.
- Most serious is aplastic anemia
 1. Parovirus (5th disease) often responsible

Anemia at School:

- ❑ Signs of an aplastic episode are pallor and lethargy.
- ❑ Identifying pallor in a child with dark skin may be difficult.
- ❑ Always consult parent if you are concerned about child's appearance or behavior.
- ❑ All fevers are taken seriously

Splenic Sequestration

- ❑ **Blood flow to the spleen is adequate, but vascular outflow from spleen to systemic circulation is occluded.**
- Life threatening
- Children under 5 years most susceptible



What to do at school:

- Because large amounts of blood is trapped in the spleen, this organ becomes enlarged.
- Child will be weak, pale, listless with a swollen and painful abdomen.
- He will perspire.
- Hemoglobin drops drastically, child can go into shock and die.
- This can be a 911 situation.

Infections

- Major cause of morbidity/mortality.**
- Susceptibility to infections is due to splenic dysfunction.**
- Spleen normal at birth, but not functional after 6 months of age.**

Infections:

Greater incidence of bacterial infections, especially pneumonia, sepsis, meningitis, osteomyelitis
a. This susceptibility to infection is related to the spleen's inability to remove bacteria from the bloodstream.

Infections

- Without a functioning spleen very susceptible to:
 1. Strep pneumonia (Pneumovax)
 2. H. influenza
 3. Neisseria meningitides (Menomune)

Infections

Importance of antibiotic prophylaxis:

Penicillin daily- usually only until age 5 or 6

Infections and School:

- Parent and/or health care facility should be notified of any temperature greater than 101°.

Chest Syndrome

Responsible for 25% of all deaths in sickle cell disease.

■ Signs and Symptoms:

1. Chest pain
2. Non-productive cough
3. Fever
4. Respiratory distress

Signs and Symptoms of Chest Syndrome:

- Chest pain
- Difficulty breathing
- Cough
- Fever
- Child will look sick

If chest syndrome occurs at school:

- Call the child's family or medical facility.
- If symptoms are severe, call 911
- Recovery is slow. Child may miss several weeks of school.
- When child returns to school, activity may need restricting for a period of time.

Neurological Problems:

- CVA (Stroke)
- TIA (Transient ischemic attack)
- "Silent strokes"

Neurological Problems: Effects on Learning:

Subtle neurological deficits related to sickling in the central nervous system vessels can impair learning and academic achievement, even without a history of neurological disease.

Learning Problems:

- Studies have shown that some children with SCD have:
 1. Lower reading and spelling scores.
 2. Do less well in visual-motor and attention skills.Scores were not related to severity of the physical illness.

Signs and Symptoms of a Stroke (CVA):

- Loss of balance
- Falls more than usual
- Weakness on one side of the body
- Headache
- Visual disturbances
- Seizure
- Academic performance decline

Preventative Measures to ~~Preserve Neurological~~ Function:

- Transcranial doppler study
- Transfusion programs

Preventative Programs:

- Chronic transfusion programs for children at high risk for a stroke or who have had one.
- Usually done every 3-4 weeks.
- Takes most of the day
- Chelation therapy- oral/IV

Renal (Kidney) Problems

- **Damage to kidney by repeated blockage of small vessels causes the kidney to lose its ability to concentrate urine.**
- **Result: children will put out large amounts of urine, even in hot weather.**

Renal Problems Result in:

- **Frequent urination**
- **Enuresis (bed wetting)**
- **Dehydration in hot weather**
- **When fluids are lost through other means: vomiting, diarrhea, perspiration, dehydration results and pain crisis can follow.**

Response in the School Setting:

- **Child must be allowed to drink and use the restroom more often than other classmates.**
- **In hot weather: excuse from strenuous outdoor activities.**
- **Remind the child to drink, especially in hot weather.**

Delay in Physical Maturation

- Some children experience physical and sexual developmental delay.
 - a. Deficits in weight is more common than in height.
- Sexual maturation for both boys and girls may be delayed by 2 years.
- Fertility appears to be normal for both boys and girls.

Helpful Hints:

- If adolescent is worried about his growth and development, or the object of teasing, it is important to acknowledge his feelings.
- Reinforce that he will catch up with his peers.
- Consider sickle cell education with peers so they can understand the disease and its effects.

Helpful Hints:

- Do not treat the adolescent who is physically immature, younger than he is.
 1. Usually, they are as emotionally and intellectually mature as their larger classmates.
 2. This is important for those striving to establish same sex relationships, as well as relationships with the opposite sex.

Other problems:

- Jaundice
- Priapism
- Gall stones

School Life

- The feeling should be fostered that a young person with sickle cell disease should be a young person first and a patient with sickle cell disease second.

- Should have as normal a school life as is possible.

Academic Performance

- If child has had a stroke or TIA there will be neurological effects.

- Poor school performance can have other sources.

Poor School Performance

- ❑ School absences.
- ❑ Child also may have subtle neurological deficits related to sickling in the central nervous system vasculature that can impair learning and academic achievement without a history of stroke or other neurological diseases.

Attendance in School

- ❑ Absenteeism will vary from child to child.
- ❑ May have many absences but the duration is never long enough to take advantage of homebound tutoring.
- ❑ Frequent absences can result in low academic skill level and behavior problems.

Attendance in School

- ❑ Self esteem cannot be fostered when forced to compete with their peers. They do not have the same information as their classmates because of missed days. Humiliation can result if they are called upon in class and cannot respond appropriately due lack of information.

Attendance in School

- ❑ Children who are frequently absent from school may be depressed and withdrawn. May be demonstrated by inattentive behaviors.

Attendance at School:

- ❑ Often overlooked is that school helps develop social skills that play a huge role in our future as adults.
- ❑ Frequent absences disrupt friendships, interfere with participation in extracurricular activities, and feeling a part of ongoing class activities.

Attendance at School:

- ❑ Teachers can help decrease absences by doing all they can to ensure that the child is not missing school unnecessarily.
 1. One study found 30% of school days missed had nothing to do with sickle cell disease.
 2. No automatic excuses. Frequent absences need a note from M.D. and contact with parents.
 3. School avoidance. Parent? Child?

Conclusions:

- Sickle cell disease is a chronic condition.
- It has the potential for many complications, some life threatening.
- The full potential of children with sickle cell disease may not be realized when home, school and medical staff are not working together to promote the health and well-being of the child.

Questions?
