



Sickle Cell Disease in the Educational Setting

School Nurse Perspective

Objectives

- Increase knowledge concerning sickle cell disease including complications and current treatments
 - Increase familiarity with issues related to working with students with sickle cell, their families and school staff
 - Address relationship of school with the sickle cell disease treatment center.
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What is Sickle Cell Disease?

Definition

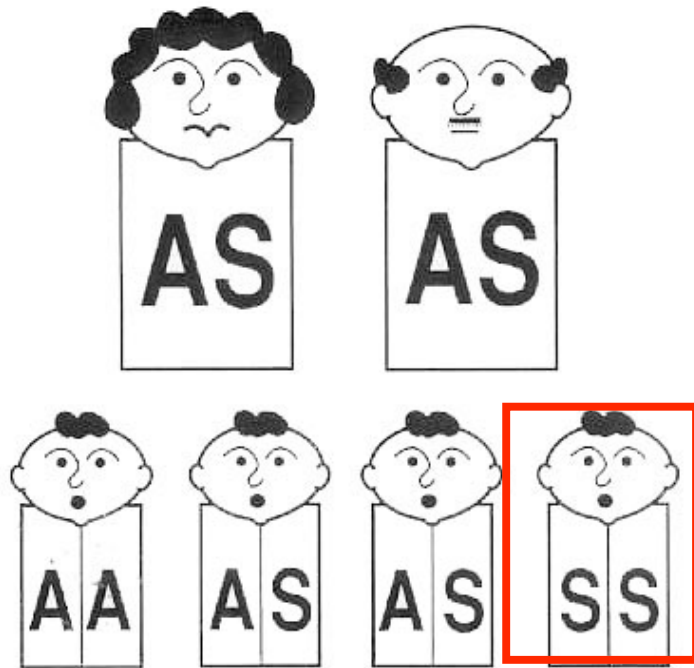
- Sickle cell disease (SCD) is a genetic disease of the red blood cell characterized by vaso-occlusion and hemolysis
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Genetic Disease: Diagnosis

- Aim of newborn screening is to identify infants with SCD to start penicillin prophylaxis



Genetic Disease: Inheritance



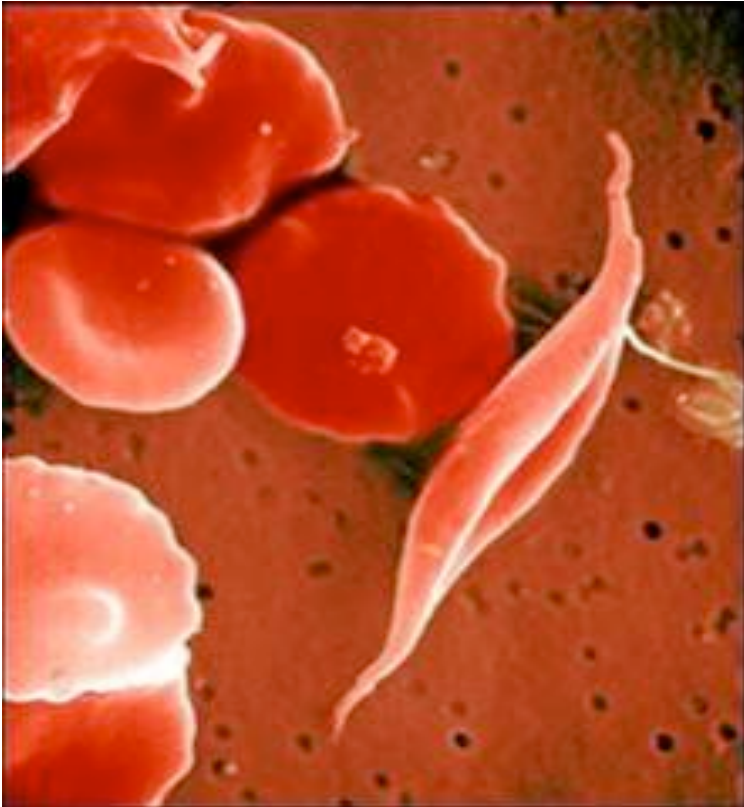
- Both parents must have an abnormal hemoglobin trait to have a child with SCD.

- If both parents have trait there is a 1 in 4 (25%) chance that each baby will have SCD.

Genetic Disease: Prognosis

- Previous studies were done before the use of hydroxyurea
 - Survival is now believed to be improved
 - Hb SS: mid-40s
 - Hb SC: mid-60s
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Red Blood Cells



- Single amino acid substitution in the gene for hemoglobin
 - Sickled cells are stiff and sharp instead of soft and round
 - Sickled cells can cause problems all over the body
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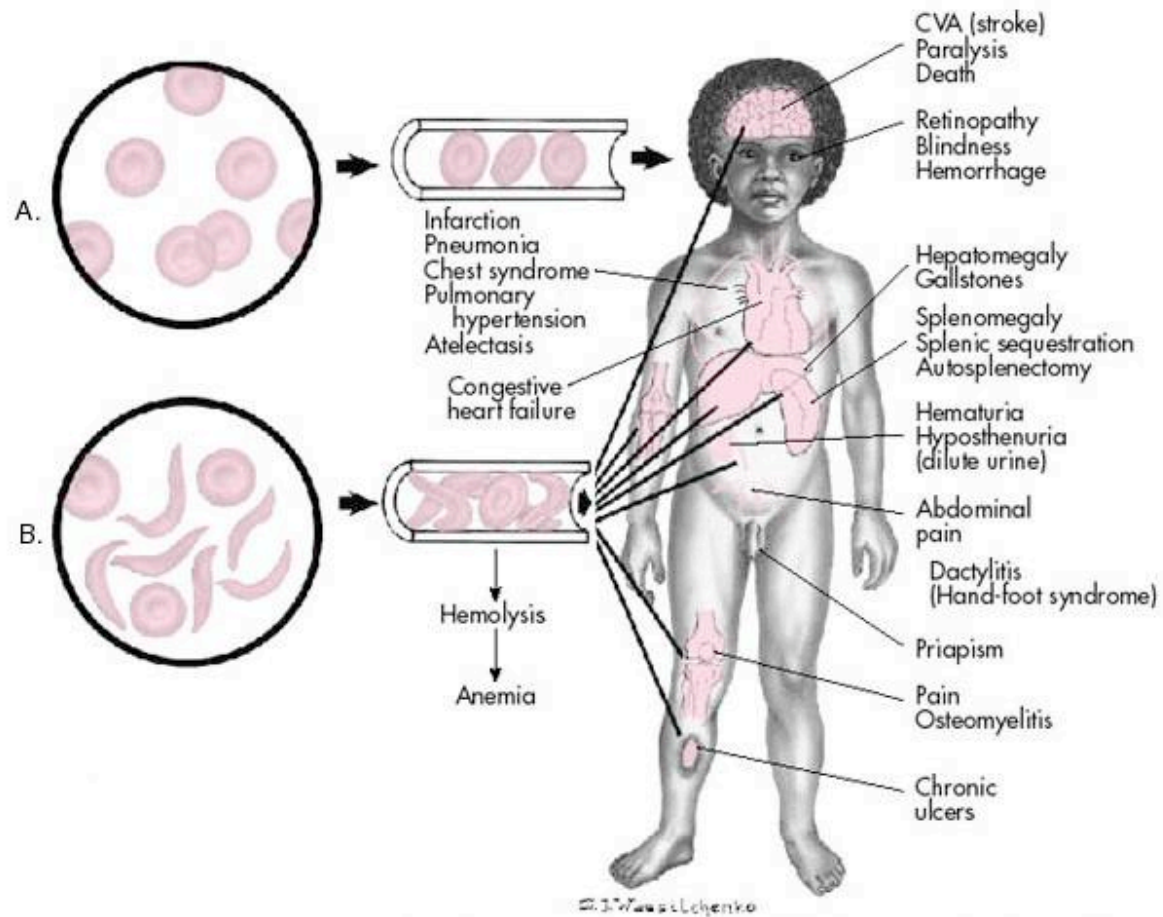
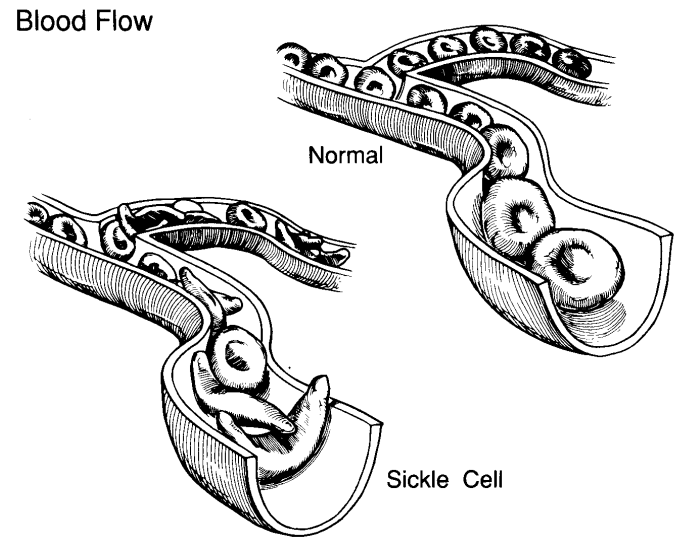


Fig. 35-4, A & B Differences between effects of, **A**, normal and, **B**, sickled RBCs on circulation with selected consequences in a child.

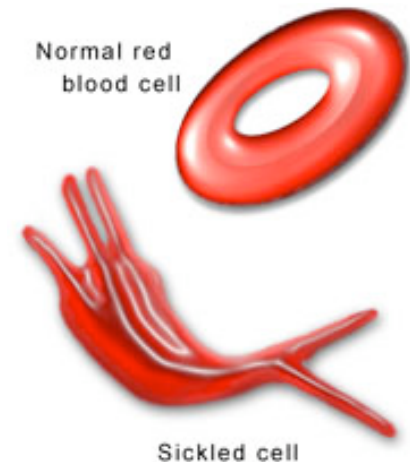
Vaso Occlusion

- All cells have the genetic defect but are not always sickled
- Sickled cells block flow within blood vessels
- Area of decreased flow is deprived of oxygen → tissue damage and pain



Hemolysis

- Normal red cell lives 120 days
- Sickled cells may only last a few days: the cells break apart increasing bilirubin levels
- Bone marrow must work harder to try and compensate



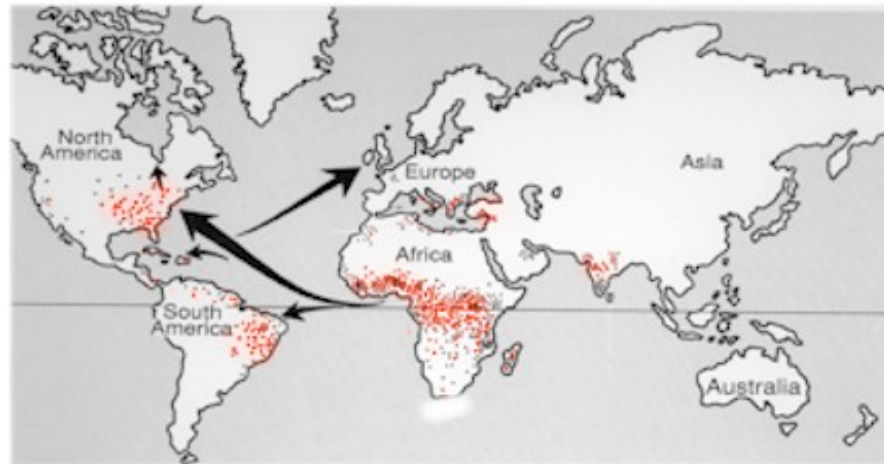
Sickle Cell Diseases

- Family of disorders: all are Hb S plus another abnormal Hb (SS, SC, SD, Sthal, etc)
- This presentation will use SCD to refer to all forms

AA	Normal (adult)
AS	Sickle cell trait
SS	Sickle cell disease: either Hb SS
SC	Sickle cell disease: HbSC
S-thal	Sickle cell disease: Hb S-beta⁺ thalassemia; HbS-beta⁰ thalassemia

Who has SCD?

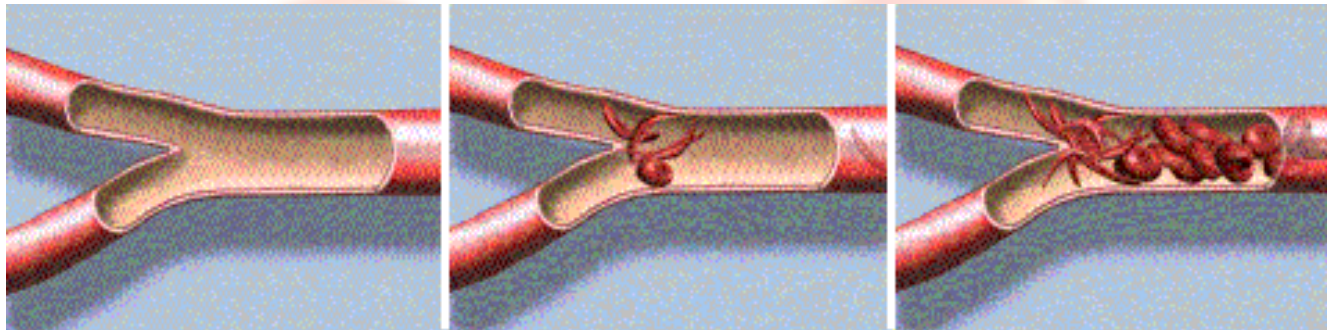
- 1 in 8 persons of African or Caribbean descent have sickle cell trait
- Also found in Hispanics, and persons from India, the Mediterranean and the Middle East



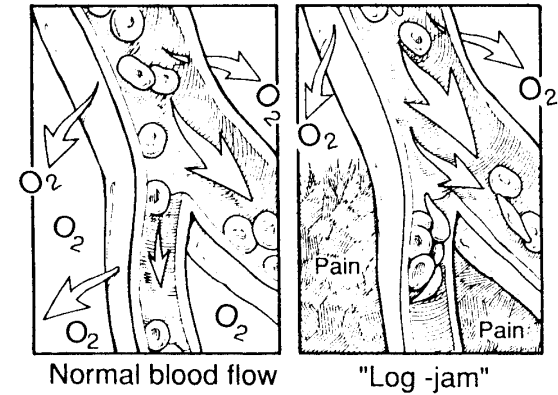
Case Study

- *Robert is a 15 y/o male with sickle cell disease in high school*
 - *Stephanie is his 7 y/o sister in elementary school*
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The Role of Vaso-Occlusion



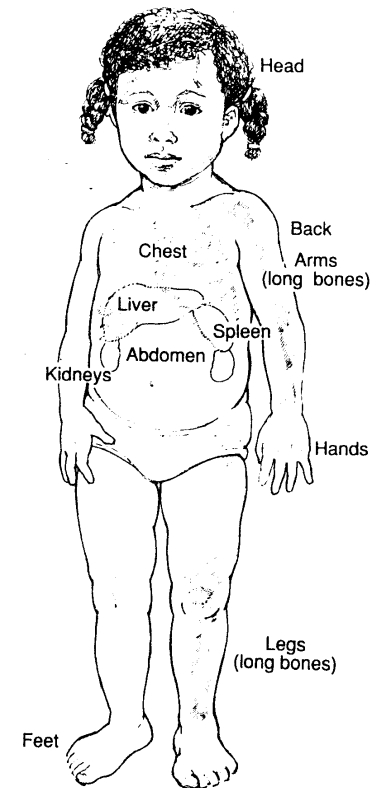
Pain



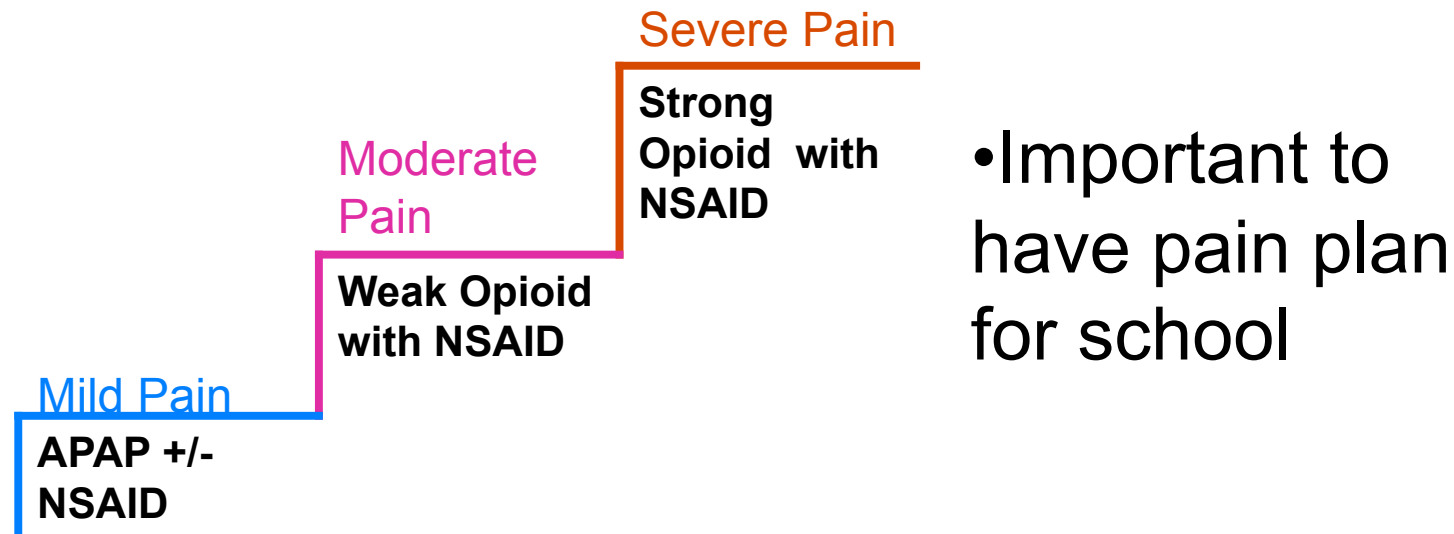
- Occlusion of small vessels leads to impaired oxygen delivery to tissues
 - Pain crises often occur in the same areas of the body
 - Older children can often discern 'sickle pain' from other pain
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Pain: Non-Pharmacologic Treatment

- Fluids, rest, warmth
- Never ice, even for sports injury
- Keep parents informed, even if child is staying at school



Pharmacologic Treatment



- Important to have pain plan for school

- Children with frequent pain crises can often stay at school, even if taking strong pain medications

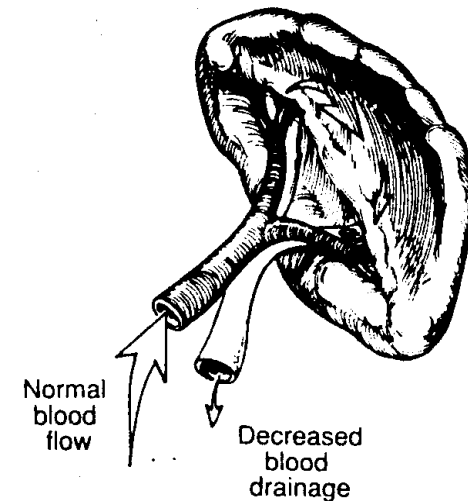
Pain: Prevention & School Concerns



- Avoid triggers: cold, getting chilled, dehydration
 - Cannot prevent all crises
 - Goal is early detection and treatment
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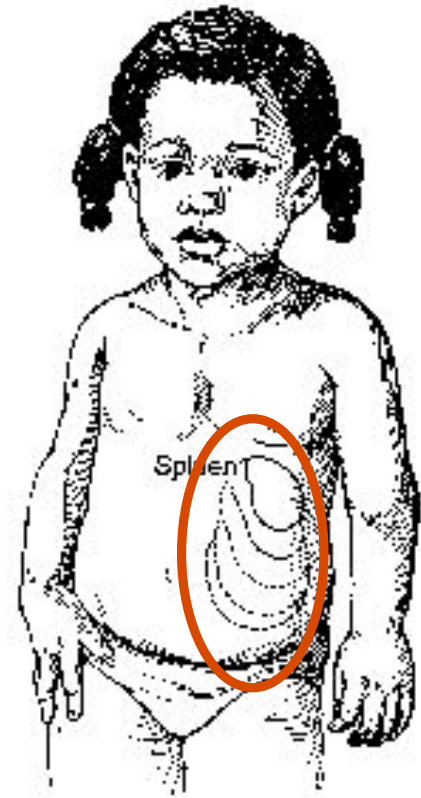
Splenic Sequestration

- Blood flows into the spleen, but does not come back out
- Causes life-threatening anemia
- Symptoms: left upper quadrant pain, pallor, fatigue, tachychardia

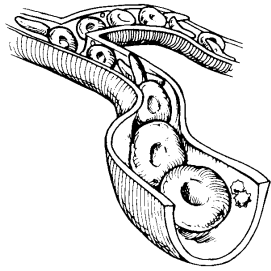


Splenic Sequestration: School Concerns (2nd slide)

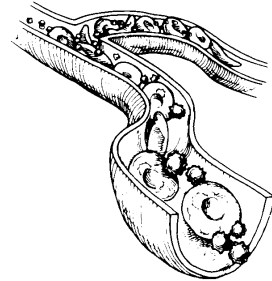
- Often recurrent
- Young children with Hb SS or Hb S-beta thalassemia
- Older children and teens with Hb SC
- Hypersplenism is risk for contact sports



Infection



Infection

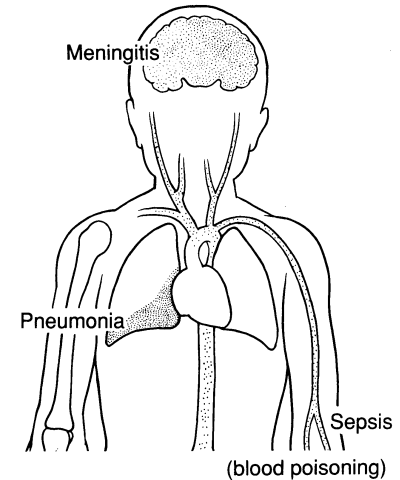


Septimia (blood poisoning)

- Sickling results in functional asplenia which may begin in infancy
 - Increased susceptibility to infections with certain bacteria
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Infection

- Any fever above 101 degrees requires evaluation and treatment
- Complications include: bacteremia/sepsis, meningitis, osteomyelitis, and pneumonia/acute chest syndrome

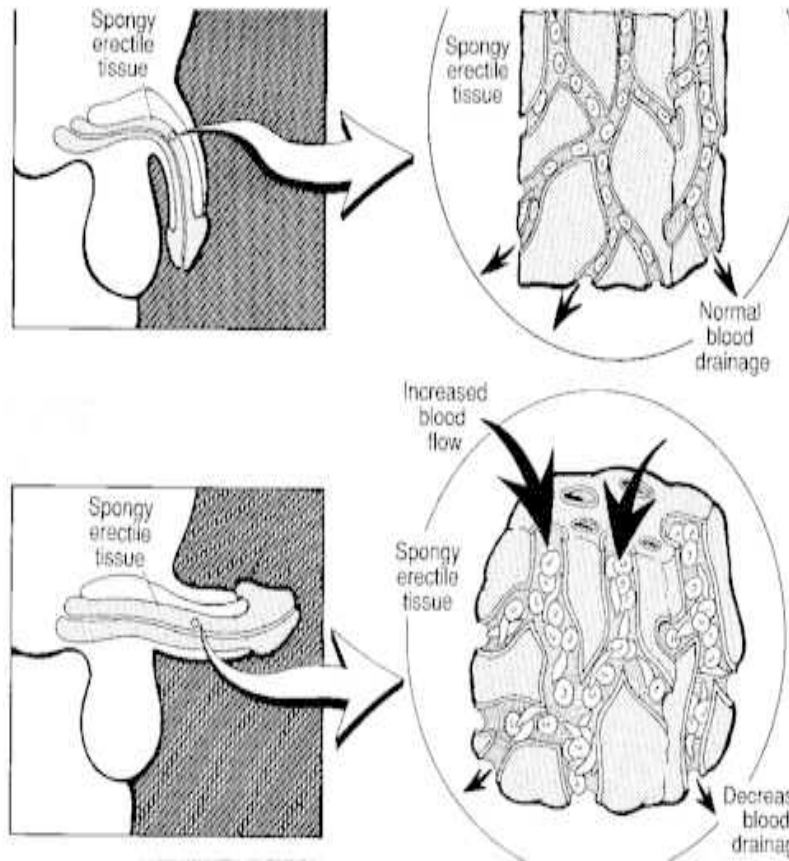


Infection: Prevention & School Concerns (2nd slide)

- Treating fever is not an option, even at family request
- Handwashing is always a good idea, especially during flu season



Priapism



- Persistent, painful, unwanted erection
 - Blood flows into the penis, but does not come back out
 - May lead to impotence
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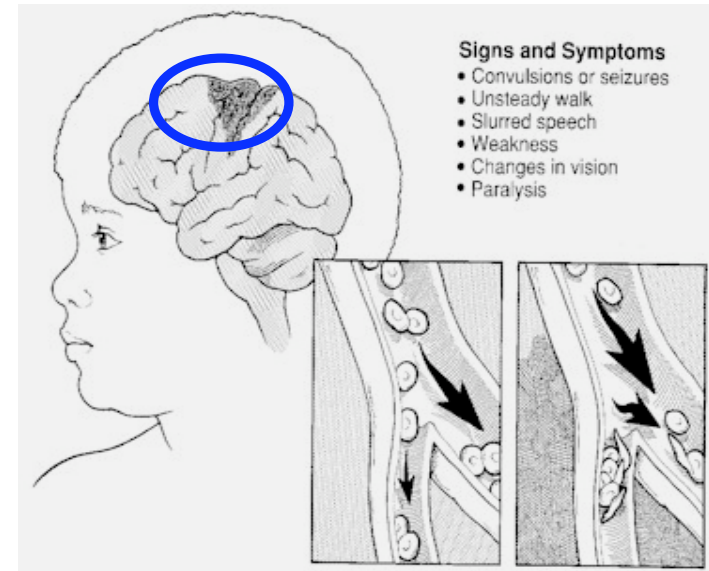
Priapism: School Concerns

- May refuse to discuss
- Treat as a pain crisis
- True priapism that lasts beyond 2 hours requires medical attention
- Treatment center should provide individualized plan



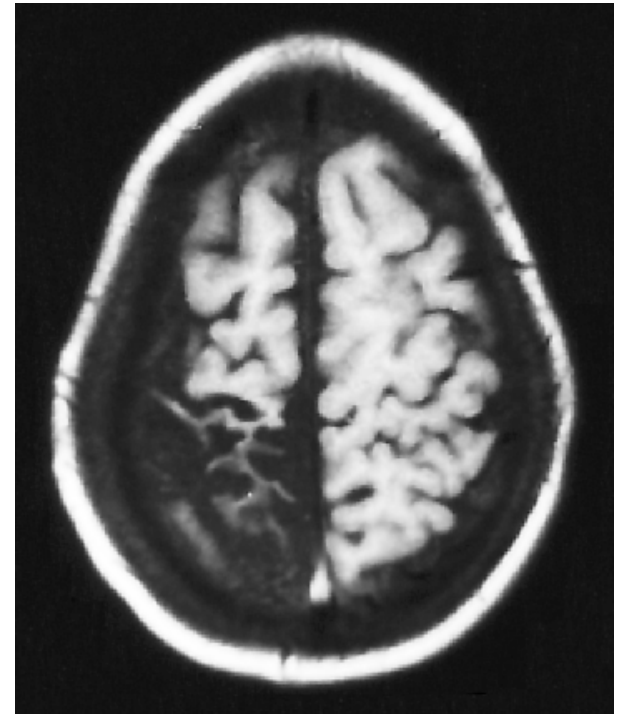
Stroke

- 10% of children with sickle cell will have a stroke, mostly those with Hb SS
- Symptoms are similar to an adult
- Prompt medical attention may reverse damage



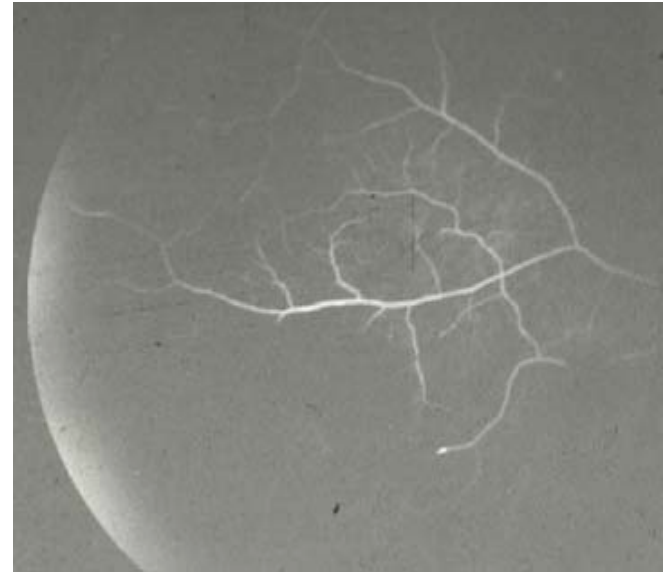
Stroke: School Concerns

- Stroke can cause subtle deficits that are not easily apparent
- Change in academic performance may be important
- Monthly transfusions can prevent recurrence



Vision Loss

- Damage to retina causes proliferative retinopathy
- Can lead to blindness
- Symptoms: seeing floaters, impaired vision



Vision Loss: Prevention & School Concerns

- Mild symptoms → significant damage
 - All children with SCD over age 10 years need annual eye exams
 - Eye injury (hyphema) is a concern for those with SCD and trait
-

Dehydration/Kidneys

- Kidneys of children with SCD are damaged and lose more fluid than normal
- Symptoms: frequent urination, increased thirst, dehydration, bedwetting
- Restricting fluids makes this worse



Dehydration and Kidneys: Prevention & School Concerns

- Free access to the bathroom and water available during class
- Exercise and heat increase fluid needs



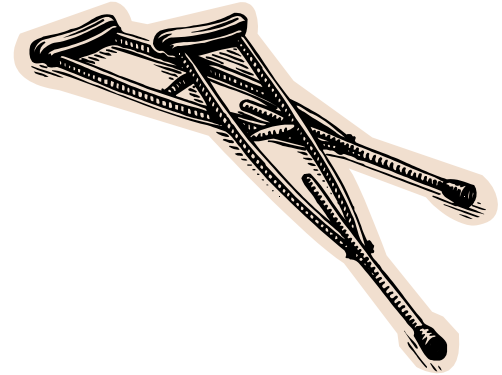
Avascular Necrosis (AVN)

- Damage to bone at the head of the femur (hip AVN) or humerus (shoulder AVN)
- Can lead to collapse and require replacement
- Symptoms: pain, limp, reduced range of motion
- Mostly older children and teens



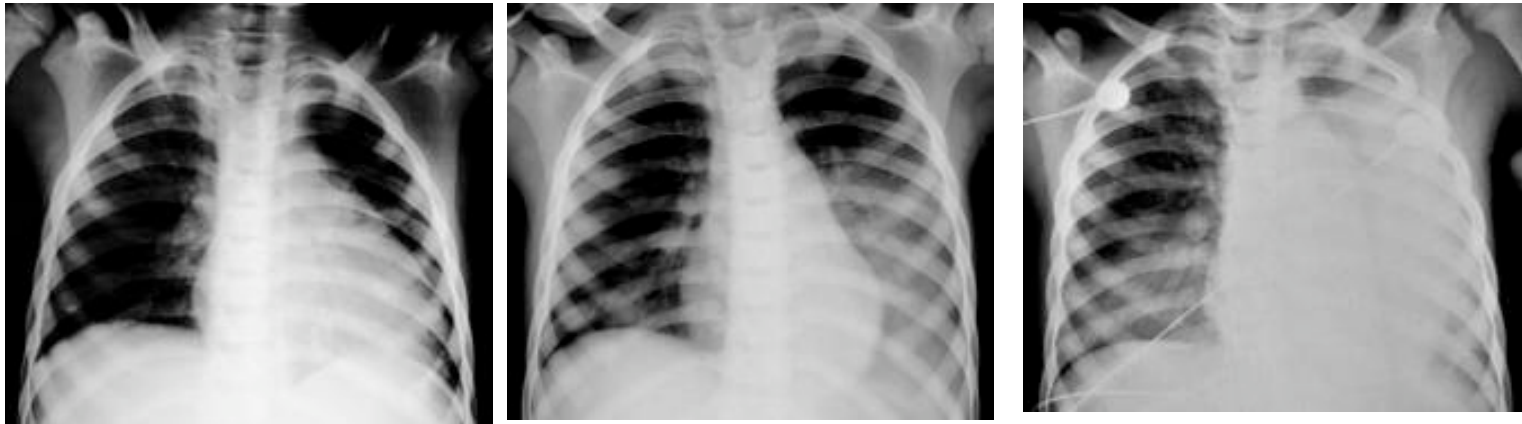
AVN: School Concerns

- Structural accommodations: elevator pass, 2nd set of books, transportation, changing classes
- NSAIDS before opiates
- May be homebound
- Encourage compliance

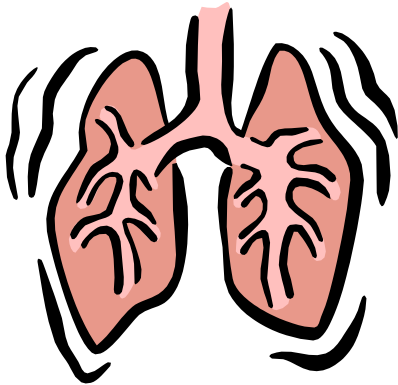


Acute Chest Syndrome

- Any change in chest x-ray with a fever or respiratory symptom
- Leading cause of illness and death
- May be infection, occlusion or embolism



Acute Chest Syndrome: Prevention & School Concerns



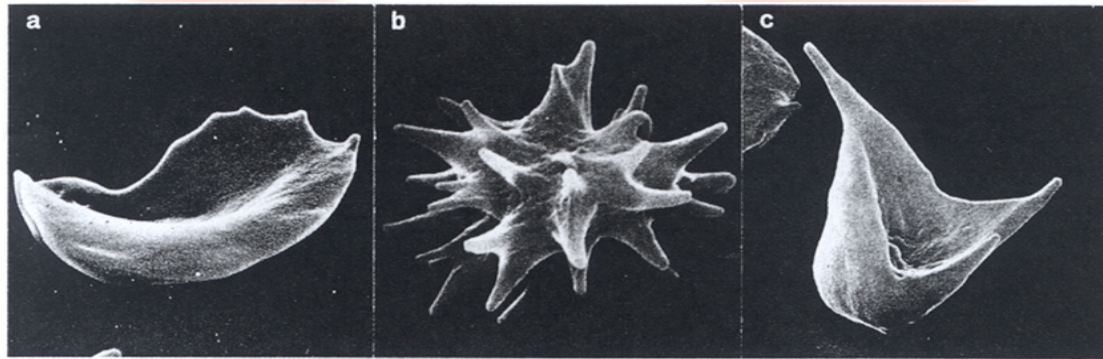
- SCD with asthma can be dangerous
- These children have more episodes of acute chest and should be taking a controller/preventative medication
- It is especially important that they have Asthma Action Plans



Case Study

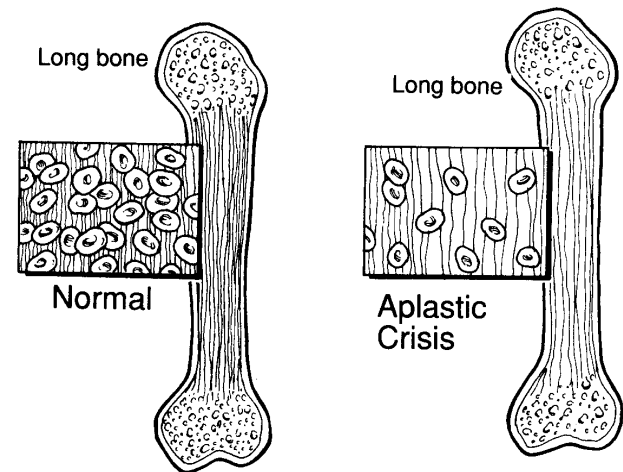
- *Robert has frequent pain crises in his back and also has AVN of both hips, he is on crutches*
 - *Stephanie had a stroke when she was in kindergarten*
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The Role of Hemolysis



Chronic Anemia

- Because sickled red cells do not live very long, the bone marrow cannot keep up
- Symptoms: benign heart murmur, fatigue
- Infection can cause severe anemia



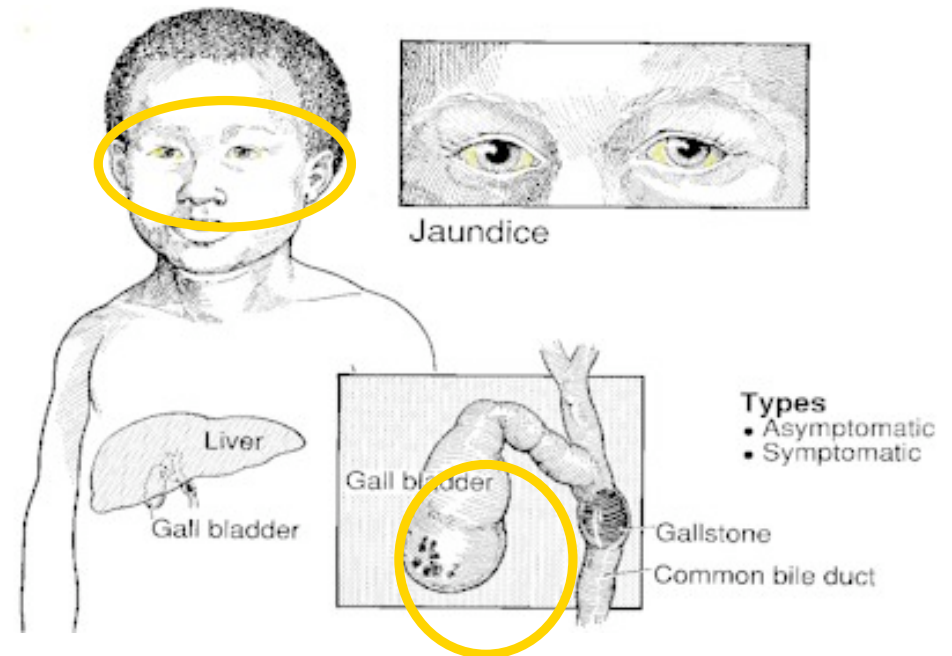
Chronic Anemia: School Concerns



- May tire easily
 - Delayed growth and development can be a source of embarrassment
 - Unusual fatigue, especially with pallor should be reported
 - Outbreaks of Parvovirus/B19
-

Jaundice & Gallstones

- Rapid red cell destruction increases bilirubin levels
- Causes scleral icterus and gallstones



Gallstones: School Concerns

- Important to know baseline icterus
- Dehydration or illness will increase jaundice
- Children may be teased because of yellow eyes
- Gallstones are not from diet



Case Study

- *Robert has gallstones and will have his gallbladder removed over summer vacation, he always has yellow eyes and is smaller than his classmates*
 - *Stephanie does not have these problems because she is chronically transfused*
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Treatments for Sickle Cell Disease

- Preventative
- Symptomatic
- Curative

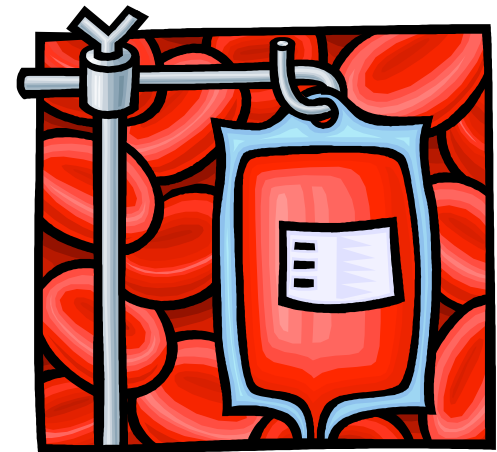
Preventative Care: Infection

- Penicillin prophylaxis until age 5 years
- Immunizations: Prevnar™, Pneumovax™ and Influenza



Preventative Care: Blood Transfusions

- Blood transfusions to prevent stroke or to protect lungs
- Why not transfuse all patients?



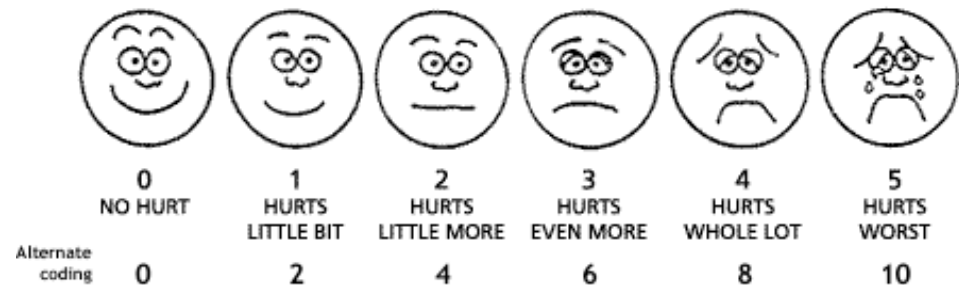
Preventative Care: Hydroxyurea

- Brand names are Hydrea™ or Droxia™
- Increases levels of fetal hemoglobin
- Red blood cells live longer and sickle less
- Can cause reversible neutropenia
- Potential carcinogen or teratogen
- Biggest problem is compliance



Symptomatic Care

- Pain medications
- Fluids
- Blood transfusions for acute illness




Cure for SCD

- Bone marrow or stem cell transplant
 - Can cure sickle cell, but risk of serious and fatal complications
 - Reserved for the sickest children with sibling matches
-

Case Study

- *Robert is starting hydroxyurea treatment and must go to clinic every two weeks*
 - *Stephanie has an implanted port for monthly transfusions, and uses Desferal infusions 5 nights a week*
-

The background of the slide is a light pink color with a pattern of semi-transparent, stylized red blood cells scattered across it. The cells are depicted in various orientations and positions, some appearing as simple circles and others as more elongated, crescent-like shapes, representing the characteristic sickle cell morphology.

Special/Recurrent Issues in Sickle Cell Care

Frequent Pain at School

- There is a small proportion of children with SCD who require frequent use of strong pain medications
- Dependence versus addiction



Strategies for Frequent Pain Medication Use

- Devise plan with family and treatment center
- Keep track of medication use
- Watch for secondary gain from trips out of the classroom



Frequent Hospitalizations or Home Illnesses

- Some children will have periods when they are unable to attend school, but do not need to be in the hospital
- Missed school plan: tutoring, assignment plan, 2nd set of books
- Must allow student to complete all required work



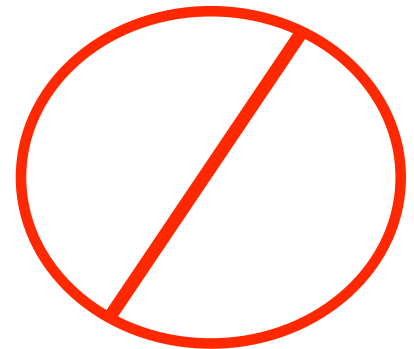
Missed School for Procedures/Transfusions

- Elective procedures are often scheduled in advance
- Encourage family to plan for missed school
- Require documentation for excessive absences



Recess, Gym and Field Trips

- Children with SCD should not be allowed to get cold, wet or chilled
- Swimming only allowed with permission of family and treatment center



Drugs and Alcohol

- Persons who take pain medications for medical reasons rarely become addicted
- Alcohol is dehydrating and can precipitate a pain crisis
- Alcohol and illicit drugs can cause fatal complications when taken with prescription pain medications



Case Study

- *Robert has several severe pain crises each year and will be hospitalized for 1-2 weeks, he also misses school on a regular basis for smaller crises handled at home*
 - *Stephanie has planned transfusions appointments and scheduled visits with specialists*
-



Collaboration

Working with Students

- Establish a relationship when well
- Encourage the early reporting of symptoms
- Teach about pain prevention
- Plan for missed school
- Expect achievement



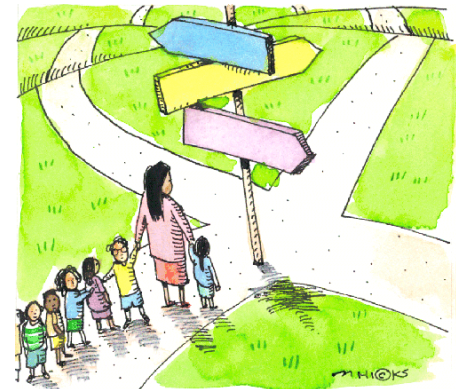
Working with Parents

- Certain families have guilt and secrecy about diagnosis
- Establish 2-way communication in the beginning of the year
- Require multiple contact numbers



Working with Parents

- Keep appropriate medications at school and require written plan
- Make a plan for missed school
- Reinforce academic expectations
- Advocate for reasonable accommodations



Supporting School Staff

- Provide information and consultation
- Clarify misconceptions
- Ensure access to RN during school
- Assist with limit setting



Supporting School Staff

- Advocate for missed school plan and other reasonable accommodations
 - Engage parents in process
 - Evaluate need for 504 plan
 - 766 referrals
-

Working with the SCD Treatment Center

- Use staff of treatment center as a resource
 - Keep center informed of concerns
 - Require written treatment plans, especially on complicated children
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Community Resources for Children and Families

- DPH Special Health Care Needs Program
 - Hole in the Wall Gang Camp
 - Next Step Program
 - STRIVE
 - Community Support Groups
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Case Study

- *Robert has a 504 plan and gets door to door transportation, a second set of books, an elevator key and has a tutoring plan*
 - *Stephanie has an IEP for her learning difficulties and her teacher provides schoolwork before her scheduled absences*
-

Conclusion

At any time, the patient with sickle cell disease can be faced with a myriad of potentially life-threatening and unpredictable complications.

Most patients and families with a good understanding of the disease process have good outcomes due to compliance with medication regimens and an overall healthy attitude that promotes well-being.

-Nedra Dodds MD, 2001



Questions?