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.
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
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
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
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

<table>
<thead>
<tr>
<th>AA</th>
<th>Normal (adult)</th>
</tr>
</thead>
<tbody>
<tr>
<td>AS</td>
<td>Sickle cell trait</td>
</tr>
<tr>
<td>SS</td>
<td>Sickle cell disease: either Hb SS</td>
</tr>
<tr>
<td>SC</td>
<td>Sickle cell disease: HbSC</td>
</tr>
<tr>
<td>S-thal</td>
<td>Sickle cell disease: Hb S-beta^+ thalassemia; HbS-beta^0 thalassemia</td>
</tr>
</tbody>
</table>
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
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
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

- **Severe Pain**
  - Strong Opioid with NSAID

- **Moderate Pain**
  - Weak Opioid with NSAID

- **Mild Pain**
  - APAP +/- NSAID

- **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
Splenic Sequestration

- Blood flows into the spleen, but does not come back out
- Causes life-threatening anemia
- Symptoms: left upper quadrant pain, pallor, fatigue, tachycardia
Splenic Sequestration: School Concerns (2\textsuperscript{nd} 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

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