SICKLE CELL DISEASE

KEYS FOR STAYING IN SCHOOL
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- Sickle Cell Disease: A group of diseases affecting the red blood cells, specifically the hemoglobin molecule. A child inherits from each parent a gene that makes an altered form of hemoglobin. This alteration in the gene causes the red blood cells to assume the sickle shape.
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DIAGNOSIS

• Hemoglobin Electrophoresis
• Newborn Screening at Birth
• Complete Blood Count (CBC), Retic Count
• Review of Blood Smear
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Types of Sickle Cell Disease  Hemoglobin
• Sickle Cell Anemia  6 - 9 grams
• Hemoglobin SC Disease  10 - 13 grams
• Sickle Beta Zero Thalassemia  6 - 9 grams
• Sickle Beta Plus Thalassemia  10 - 13 grams
• Sickle Cell Trait  Normal
Sickle Cell Disease
Keys For Staying In School

Nursing Assessment

a. Eye - retinopathy, vision problems
b. Blood - anemia
c. CV - heart murmur
d. Endocrine - delayed growth and development
Sickle Cell Disease
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Nursing Assessment

e. Musculoskeletal - Avascular Necrosis of Femoral Head/Humerus; bone/joint pain, limping

f. Hepatobiliary - Jaundice, gallstones, spleen/liver sequestration

g. Renal - enuresis, kidney damage
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Complications and Nursing Interventions

A. Pain

Causes: Vaso-occlusion of Sickled Cells
Goal: Control pain and side effects of pain medications.
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Treatment: Hydration, opioids, NSAIDS, Rest

Nursing Intervention: Alternate opioids every 2 hours with NSAIDS every 2 hours. Give different medication every 4 hours. Rest in office for 1 to 2 hours.
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B. Fever/Infection Risk

Cause: Absence of splenic function
Goal: Prevent sepsis or blood infection
Treatment: Prophylactic antibiotics, fever precautions, IV antibiotics, blood cultures
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Nursing Intervention: Evaluation by health care provider for temperature greater than or equal to 101.5 degrees.

Immunization Recommendations:
- Pneumococcal (Prevnar, Pneumovax)
- HIB, Menactra
- Influenza, H1N1
C. Stroke

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Goal: Decrease stroke risk or recurrence by suppression of red blood cell production.

Treatment: Monthly transfusions with packed red blood cells.
Nursing Intervention: Medical Emergency if symptoms occur. Dial 911. Use emergency procedures if necessary. Post stroke interventions include safety risks, assessment of class placement, assistance with physical challenges, i.e., crutches, wheelchairs, etc.
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D. Surgery
Causes: Removal of organ/body part due to poor function, resulting from chronic organ damage from sickle cells.

Common Surgeries:
1. T&A: overgrowth of tonsillar tissue, sleep apnea, snoring
2. Splenectomy: spleen damage, pain in left upper quadrant, jaundice, pallor
Common Surgeries:

3. Cholecystectomy: gallstones; pain in right upper quadrant, nausea and vomiting after fatty meals, jaundice

4. Hip Core Decompression: damage to heads of femur and humerus; pain in hips and knees when using stairs; limping
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Goal: Relieve symptoms related to poor organ function.
Treatment: Surgical intervention, if necessary, to relieve symptoms.
Nursing Interventions: Support after post-operative period and return to school.
E. Exercise and Physical Activity
Cause: Decrease stamina and exercise tolerance due to anemia and poor oxygen capacity.

Goal: Maintain adequate exercise tolerance during activity.
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Treatment: Participation in sport activity as tolerated and as safety permits. Monitor for overexertion and dehydration.

Nursing Interventions: Recommend or substitute alternative classes; allow for frequent rest, water and bathroom breaks; discuss limitations with coaches.
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SCHOOL PROGRAMS

A. Other Health Impaired: Allows for special accommodations and modifications in curriculum and content. Example: stroke, learning difficulties
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SCHOOL PROGRAMS

B. 504 Program: Provides access to school services. Recommended for all students with chronic illnesses. Example: Children needing rest and bathroom breaks, water privileges
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SCHOOL PROGRAMS

C. Homebound: Home instruction if out of school a cumulative total of 4 weeks or more. Instruction time 4 hours per week. Initiated by school nurse. Example: Child with Sickle Cell needing Bone Marrow Transplant.
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D. Admission, Review and Dismissal (ARD) Assessment for special education services. Development of Individual Education Plan. Allows written plan of care. Participants may include members of hospital team. Example: Stoke, ADHD, Learning Disabled
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SCHOOL PROGRAMS

SICKLE CELL DISEASE KEYS FOR STAYING IN SCHOOL

THE SICK ROLE
Requires an evaluation by a mental health professional. Child may report symptoms during activities they wish to avoid. Child may report symptoms to gain the special attention usually received during sick episodes. Usually considered after medical reasons have been ruled out.
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CHILDREN’S MEDICAL CENTER, DALLAS STAFF RESOURCES

Bonita Conley, RN, PNP-BC
  Pediatric Nurse Practitioner
Mary Lazarus, Psy.D  Psychologist
Kelly Ihejiawu, M.ED  School Liaison