Care of School Age Children with Sickle Cell Disease

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Sickle cell disease around the world
1/12 African Americans carry Sickle cell trait
1/4 West Africans carry Sickle cell trait
1/100 – 1/10 Central Asians carry Sickle cell trait

1/400 African Americans have Sickle Cell disease
1/1000 Hispanic Americans have Sickle Cell disease

~ 70,000 Americans have Sickle Cell disease
Types of Sickle Cell Disease

- SS
- SC
- SB+thal
- SB0thal
- SD
- SE
Normal red blood cell

Red blood cell with hemoglobin A

Sickle cell

Red blood cell with hemoglobin S
The Sickling problem

- **Hgb A with O2**
  - RBC is round

- **Hgb A without O2**
  - RBC is round

- **Hgb S with O2**
  - RBC is round

- **Hgb S without O2**
  - Hgb S stick together to form a rod, the rods come together to form a cable, which kinks the red blood cell into a **rigid** sickle shape.

What are some of the complications of sickle cell disease?

- Fatigue
- Jaundice
- Back Pain
- Arm and Leg Pain
- Breathlessness
- Hand Swelling
- Joint pain
- Foot swelling
Types of pain

**Acute sickle cell pain crisis** - minutes to days, caused by blocked blood flow from the sickle RBCs. Usually deep in the bones and muscles of the arms, legs, and back.

**Chronic pain** from sickle cell bone damage (avascular necrosis, osteonecrosis) - lasts longer than a few weeks and may be present constantly, due to bones being damaged by blocked blood flow. Often in the hips.

Hydroxyurea prevents pain!
Treatment for pain

**Ibuprofen = Motrin = Advil.** Blocks pain in muscles and bones. Does not make you drowsy. Can cause stomach upset and ulcer, and should be taken with food in the stomach. Good for menstrual cramps.

**Lortab** = hydrocodone + tylenol.

**MS Contin** = long-acting morphine. Best for chronic pain.

**MS IR** = immediate release morphine. Good for breakthrough pain, if already on long acting morphine.

**Methadone** - a form of morphine that is not addicting. Good for chronic pain.
Fatigue

Individual with sickle cell disease

Sickle cells break open (hemolysis)

Anemia (not enough RBCs)

Low energy

Sickle cells creating blockage

Hydroxyurea: increases hemoglobin F, prevents sickle cells from breaking down

Folic acid: helps make more red blood cells

More RBCs

More energy
Stroke

= blocked blood flow in the brain

10% of sickle cell patients have stroke
**Signs of Acute Chest Syndrome:**

- Fever
- Labored breathing
- Wheezing
- Chest pain

**Treatment:** Oxygen, antibiotics, breathing treatments, transfusion
Prevention of Acute Chest Syndrome (ACS)

Hydroxyurea !

Albuterol & Flovent inhaler !
Pain prevention and management

**F** - for Fluids and Fever. Drink lots of water. Carry a water bottle.
If you get a fever, see your doctor.

**A** - for Air. Get enough oxygen. No airplanes with unpressurized cabins, commercial airplanes OK.

**R** - for Rest. Do not overdo it. Take plenty of breaks when your body feels tired.

**M** - for preventive Medicines. **Hydroxyurea** for pain prevention. Folic acid for making new red blood cells. Asthma meds to prevent asthma attack and chest pain.

**S** - for Situations to avoid, like too hot or too cold. Avoid smoking, alcohol, or illegal drugs. Avoid emotional stress by pacing projects and work. Join a support group.
# What School Nurses need to know about Sickle Cell Disease

<table>
<thead>
<tr>
<th>Assessment</th>
<th>Observations</th>
<th>Recommendations</th>
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<tbody>
<tr>
<td>1. Pain Event</td>
<td>Complaint of arms, legs, back, or</td>
<td>Rest, notify parent. Administer medication according to medical provider's approval</td>
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<tr>
<td></td>
<td>abdomen hurting</td>
<td></td>
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<tr>
<td>2. Fever</td>
<td>Temp 100.4 or higher</td>
<td>Notify parent</td>
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<tr>
<td>3. Hot Weather</td>
<td>Temperature &gt; 90 degrees</td>
<td>Dress lightly, increase fluids, cool area or air conditioned area, limited outside activity. Increase frequency of water and bathroom breaks.</td>
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<tr>
<td>4. Cold Weather</td>
<td>Temperature &lt; 40 degrees</td>
<td>Dress warmly, hat, gloves and boots in the snow. No outside activities</td>
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<td>5. Fatigue</td>
<td>Decreased activity, decreased class participation, sleeping in class</td>
<td>Rest, limitation of physical activity including gym</td>
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<tr>
<td>6. Nose bleeds</td>
<td>Bleeding continues without intervention</td>
<td>Apply pressure for 20 min. do not apply ice</td>
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<tr>
<td>7. Decrease school performance</td>
<td>Gradual or abrupt change in behavior or academics</td>
<td>Check hearing, vision. Notify parent, may require testing, IEP</td>
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<td>8. Slurred speech, limp or headache, decrease concentration</td>
<td>Change in physical or cognitive ability</td>
<td>May signal impending stroke. Notify parent. If no response after 30 min call ambulance.</td>
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<td>9. Growth and Development</td>
<td>May be smaller than other peers( but not necessarily)</td>
<td>Will mature at a slower rate</td>
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<tr>
<td>10. Other injuries</td>
<td>Any injury to extremities, hit in eye</td>
<td>DO NOT apply ice to area, ok to use heating pad. Notify parent</td>
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Famous people with Sickle Cell Disease

PAUL WILLIAMS, THE TEMPTATIONS

Miles Davis

Georgeanna Tillman, the Marvelettes
ANY QUESTIONS?