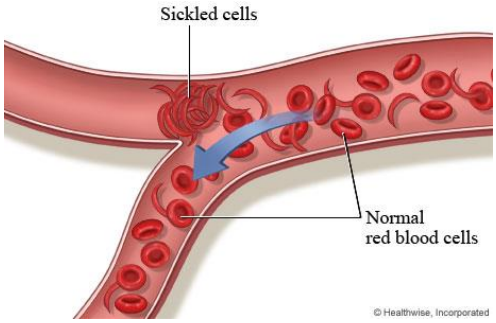
Sickle Cell Disease (SCD)
For Healthcare Providers

This is a customized health care provider version of our website. Please visit the main website to find more comprehensive information for families and schools (www.gemssforschools.org).

Physical characteristics and/or symptoms

Note: not all people with SCD will have all of these features.

Sickle cell disease (SCD) is an inherited blood condition that is characterized by pain crises and anemia. The diagnosis of SCD is made by testing for the presence of significant quantities of **Hemoglobin S** (Hb S). The severity of the disease can vary widely between individuals. Supportive treatment is available, but currently there is not a cure. Children with sickle cell disease do not have any distinct physical features.



- Hb S causes the red blood cells to become shaped like a sickle or a crescent. The symptoms of SCD are caused by these sickle-shaped red blood cells.
- When the red blood cells become sickled, they break down prematurely leading to anemia.
  Normal red blood cells are smooth and can change their shape to flow though the blood vessels. The sickle shaped cell is very rigid. The cells become trapped and "jam" blood vessels. They interfere with normal blood flow. Pain can result.
- Certain physical and environmental conditions may trigger the red blood cells to become shaped like a sickle or crescent. These include:
  - Physical exertion
  - Fever or illness

- Weather changes
- High elevation
- Swimming in cold water
- Exposure to cold
- Dehydration
- Trauma
- Emotional stress
- Unknown factors

Children with SCD need 24-hour access to medical facility that can provide urgent evaluation and treatment of any acute illness. Hydration and anti-inflammatory agents and pain medications (nonsteroidal anti-inflammatory and narcotic analgesia) are helpful.
Hydration is very important and children with sickle cell disease should be allowed unrestricted access to drinking water. Caffeine causes blood vessels to restrict so should be avoided.

- **Pain**
  - Pain is caused by sickled red blood cells, which are stiff and inflexible. They get stuck in the small blood vessels and become trapped causing "log jams" inside a blood vessel. This leads to tissues and organs being deprived of oxygen rich blood and poor blood flow, it can cause pain and sometimes swelling. Typically, younger children often complain of pain in extremities. Older individuals more commonly experience pain in the head, chest, abdomen, and back.
- **Anemia**
  - Anemia can lead to shortness of breath, fatigue, and delayed growth and sexual development in children. The rapid breakdown of red blood cells can lead to jaundice.

- **Dactylitis** is one of the earliest signs of SCD in infants and children.
  - Dactylitis is pain and/or swelling of the hands or feet, sometime referred to as "hand-foot syndrome."

- **Restricted blood flow**
  - Sickling events lead to ischemia and variable degrees of the destruction of red blood cells. This can lead to multi-organ damage and chronic pain. Organs can include: bones, lungs, liver, kidneys, brain, eyes, and joints.

- **Spleen problems**
  - When sickle cells are trapped in the blood vessels inside and leading out of the spleen, the normal flow of blood is blocked. This can lead to the spleen being engorged or over-filled with blood cells
- **Priapism**
  - This is painful and unwanted erections, which often start in childhood and often, occur in the early morning hours.

- **Infections**
    - Septicemia
    - Meningitis with pneumococcal and other bacteria
    - Pneumonia
    - Osteomyelitis
    - The most common cause of death in children with SCD is Streptococcus pneumonia sepsis. Death risk highest in the first few years of life.
    - Most children with SCD are vaccinated against these organisms and begun on prophylactic penicillin, which decreases the incidence of infections.

- **Lung issues**
    - Acute chest syndrome is caused by red blood cells trapped in the lungs. Signs of acute chest syndrome include chest pain, fast breathing and or retractions, congested "pneumonia-like cough," abdominal pain, fever, trouble breathing.

- **Aplastic Crisis**
    - Aplastic crisis causes red blood cell (RBC) production to be shut down for 10 days. Signs of aplastic crisis include; paleness, lethargy, not feeling good, headache, fever, low blood count, recent respiratory infection, and passing out.

- **Stroke**
    - Presenting signs and symptoms include; severe headaches, marked dizziness, visual changes, acute onset of weakness in a limb or one side of face, sudden inability to produce speech, or a seizure. It occurs more often in children than adults. The peak occurrence is between 2-9 years. The most common cause of stroke in children is cerebral infarction or a blockage of oxygen supply to the brain by sickled cells.

Recommended Routine Surveillance

- Routine medical and special evaluation
- Yearly for healthy affected individuals; CBC and reticulocytes count, assessment of iron status, liver and renal function tests, urinalysis, LDH, and vitamin D levels. Extended red cell phenotyping should e done at least once
- Starting at age 7 years evaluation for end-organ damage evaluation for chest x ray, ECG, pulmonary function tests, abdominal ultrasound and ophthalmology evaluation
- Individuals with Hb SS and HB SB thalassemia yearly transcranial Doppler to determine the risk of stroke
- Cardiac and pulmonary evaluations including echocardiogram, pulmonary function tests and sleep studied
- Mental health and neurocognitive assessment
- Routine dental care

Emergency Protocols

- CDC: Tips for Supporting Students with Sickle Cell Disease https://www.cdc.gov/ncbddd/sicklecell/documents/tipsheet_supporting_students_with_scd.pdf
- An emergency action plan should be in place that has information helpful to the school and to coaches and other personnel. See the previous section for examples.
- A plan to deal with mild-to moderate episodes of pain should be in place. The main treatment for pain episode is supportive. Pain episodes are managed with a multi-model approach (i.e. warmth, massage, distraction, acupuncture, bio-feedback, and self-hypnosis).
- School Personnel should realize that prophylactic antibiotics (including penicillin in children), may need to be taken at school even when no infection is present.
- Children who have SCD should have access to the nurse's office as requested.
- Be aware of signs of infection. Any infection in a child who has SCD is an emergency. Prompt evaluation and treatment of underlying infections is essential. Symptoms might include: fever, coughing, vomiting, diarrhea, crankiness, rapid breathing, pale color, unusual sleepiness, trouble breathing.
- Avoid dehydration, extremes of temperature, physical exhaustion, and extremely high altitude

Specialists Who May Be Involved

Follow up is need on a case-by-case basis. A multidisciplinary team approach to best meet the child's individual needs is recommended.

- Cardiologist
  - Tricuspid regurgitant jet
  - Cardiomyopathy
- Geneticist/genetic counselor
  - Diagnosis
  - Coordination of care
  - Genetic risk for family
  - Clinical trials
- Hematologist
  - Anemia
  - Hemolysis
  - Splenic sequestration
  - Aplastic crisis
- Neurological
  - Strokes
  - Silent cerebral infarcts
  - Cerebral hemorrhage
  - Cerebral blood flow abnormalities

    - Cerebral microvascular disease
- Neurocognitive evaluation
- Ophthalmologist
    - Retinopathy
- Pulmonologist
    - Pulmonary hypertension
    - Lung disease
    - Acute chest syndrome
- Urologist
    - Priapism

Sample Forms

- Sample paragraph to be used for Letters of Medical Necessity or Letters to the school:

My patient_____ has been diagnosed sickle cell disease (SCD). This condition is characterized by chronic anemia and acute events related to lack of blood flow. Medical complications with SCD included acute and chronic pain as well as damage to the spleen, resulting in sensitivity to infections, and decreased blood flow to the brain, which can result in stroke. The acute events can be brought on by physical stress, dehydration, changes in temperature and changes in altitude. Therefore, it is vitally important that my patient avoid dehydration, extremes in temperature, physical exhaustion, and high altitude. Because of these, _____ needs the following accommodations.

- Examples of Emergency Action Plans

https://il01001099.schoolwires.net/cms/lib/IL01001099/Centricity/Domain/281/SCAactionplan%20pdf.pdf

http://health.usf.edu/~/media/Files/Medicine/Orthopaedic/Sickle%20Cell/SCTEAP.ashx?la=en

Seven Important Aspects of School Life

"Sickle Cell Disease at a Glance" will help you talk with parents and schools about:

- Medical / Dietary Needs
- Education Supports
- Behavior & Sensory Supports
- Physical Activity, Trips, Events
- School Absences & Fatigue
- Emergency Planning
- Transitions

Resources

GeneReviews Sickle Cell Disease
http://www.ncbi.nlm.nih.gov/books/NBK1377/

American Academy of Pediatrics (AAP) – Health Supervision Guidelines
http://pediatrics.aappublications.org/content/109/3/526

National Heart, Lung, Blood Institute: evidence-based management of Sickle cell disease: expert Panel Report

https://www.nhlbi.nih.gov/health-pro/guidelines/sickle-cell-disease-guidelines/

Genetics Home Reference

https://ghr.nlm.nih.gov/condition/sickle-cell-disease

Additional resources including support group information can be found on the main website.