# Management of Sickle Cell Disease in the Educational Setting

Prepared by
Barbara Obst MSEd, BSN, RN, NCSN
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We are all born with great potential.
Shouldn't we all have the chance to achieve it?

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# Purpose

To provide a training tool for school personnel to understand the management of sickle cell disease (SCD) in the educational setting.

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# Objectives

- Increase knowledge about SCD, including complications and current treatments.
- Increase knowledge base of school personnel related to potential issues students with SCD could experience.
- Discuss potential accommodations in the school for the students with SCD.
- Discuss the usage of SCD Emergency Care Plan in the school setting.

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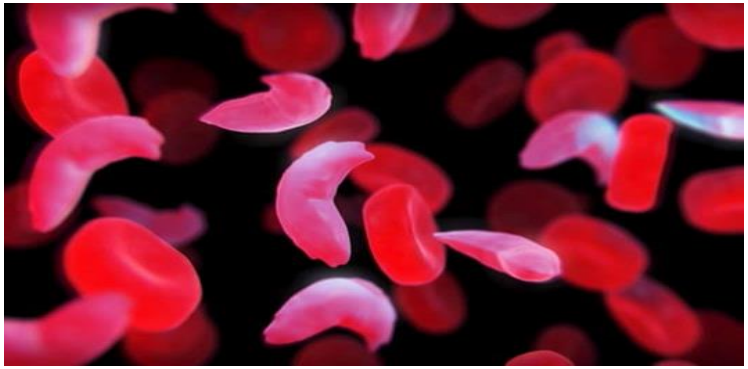
# What is Sickle Cell Disease?

- SCD is a collective term used to describe a spectrum of genetic disorders that affect the shape and function of the red blood cell (RBC's).
- RBC's form crescent or sickle-shaped cells that slow or block blood flow.



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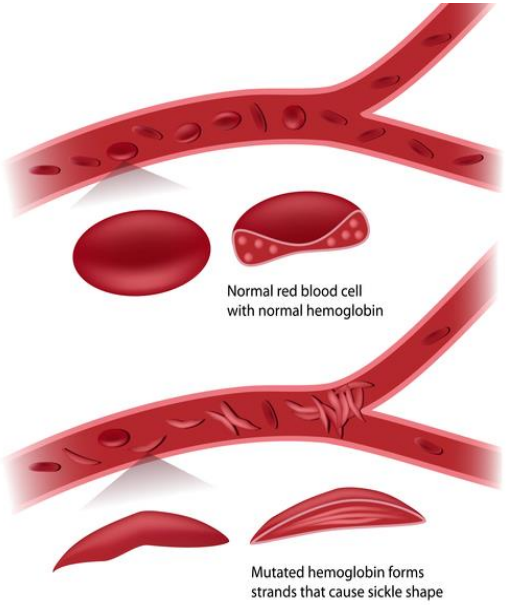
- People with SCD produce an abnormal type of hemoglobin (Hemoglobin S). Hemoglobin is a protein that carries oxygen from the lungs throughout the body.
- The abnormal hemoglobin in SCD can cause the red blood cells to have a sickled or banana shape under certain conditions.
  - With this shape, RBC's can become inflexible and sticky, causing them to adhere to vessel walls and clog blood flow.
  - Oxygen then cannot reach the intended target. Without proper tissue oxygenation, the child can experience sudden, severe pain as well as other complications.

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# About SCD

- SCD is the most common genetic disorder identified in African Americans. SCD is also found in people from South and Central America, the Mediterranean, and the Middle East.
- The three most common types of sickle cell disease are hemoglobin:
    - SS Disease
    - Hemoglobin SCD
    - Sickle Beta-Thalassemia.
- There is no universal cure for SCD.

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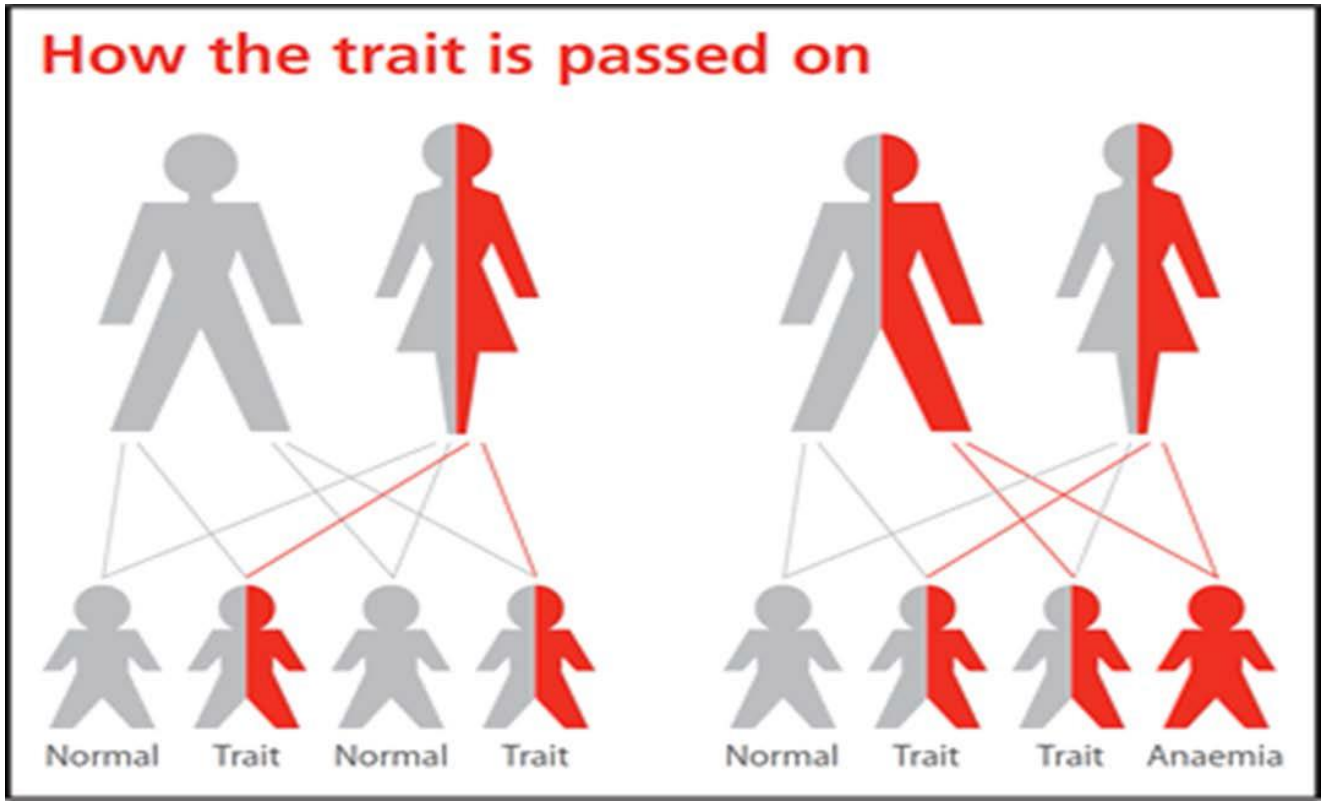
SCD is an inherited blood disorder.



# Symptoms

The severity and presentation of signs and symptoms can vary widely from person to person and change over time.

| | |
|---|---|
| Pain (bone, joint, hip) | Swelling of hands and feet |
| Headache | Weakness |
| Joint pain | Shortness of breath |
| Fatigue | Frequent urination |
| Abdominal pain | Perspiration |
| Vision problems | Anemia |
| Jaundice | Fussiness |
| Frequent infection | Delayed growth |
| Anemia | Coldness in extremities |

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# Symptoms requiring immediate action

Other symptoms should be considered **warning signs** that require immediate action and *should be outlined in the student's emergency care plan.*

| | |
|---|---|
| Fever | Change in alertness/confusion |
| Rapid breathing | Difficulty breathing |
| Rapid heartbeat | Sudden onset, blurry vision |
| Muscular weakness | Sudden onset pain |
| Sudden onset headache | Inability to speak |
| Change in complexion | Stomach pain or swelling |

Acute illness characteristics of common childhood illnesses such as fever, cough, abdominal pain, pallor, and extremity weakness, may rapidly progress to life-threatening complications.

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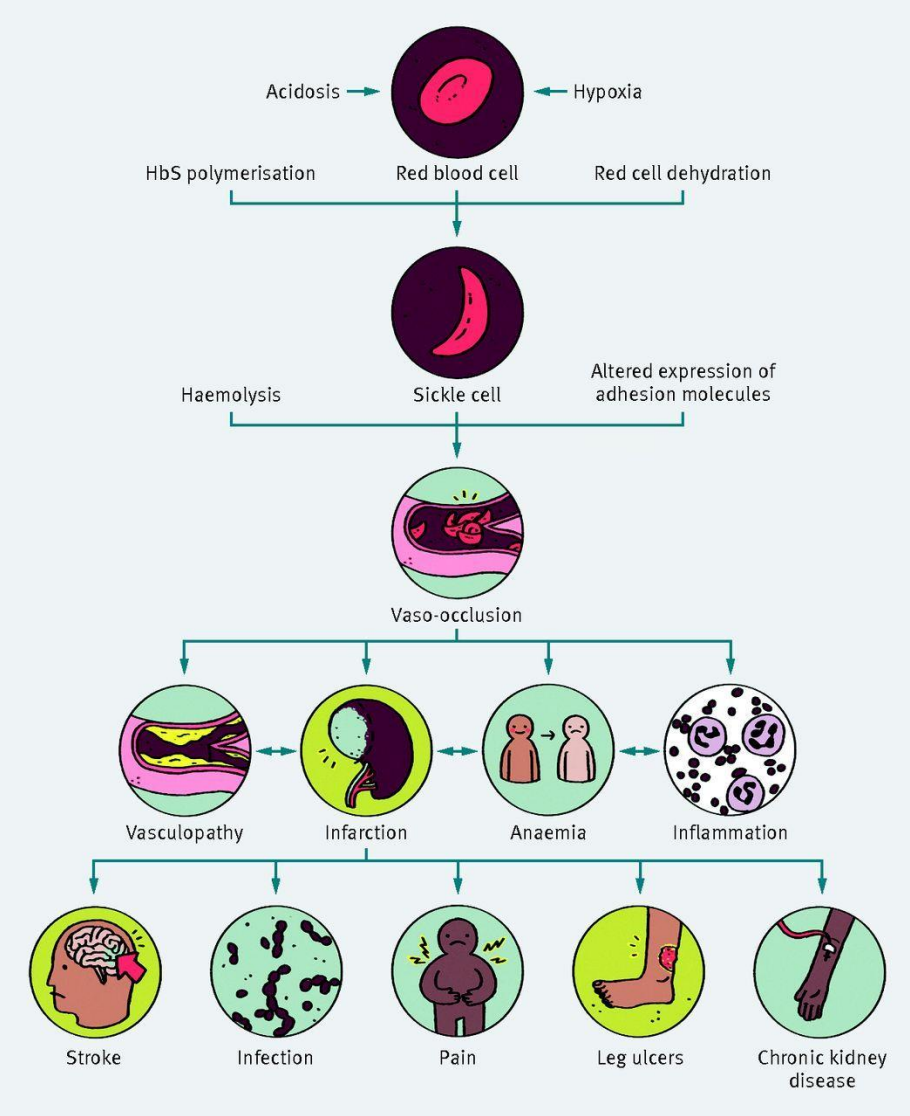


Potential complications caused by VOC.

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# Vaso-occlusive crisis (VOC) or “Pain crisis”

Pain is the hallmark symptom of the disease.

- Pain can have a sudden onset, vary in intensity from mild to severe, and last for hours to weeks.
- Can occur without warning when sickle cells block blood flow and decrease oxygen delivered to an organ.
- Acute episodes of severe pain “crises” are the primary reason that these patients seek medical care in hospital emergency departments.

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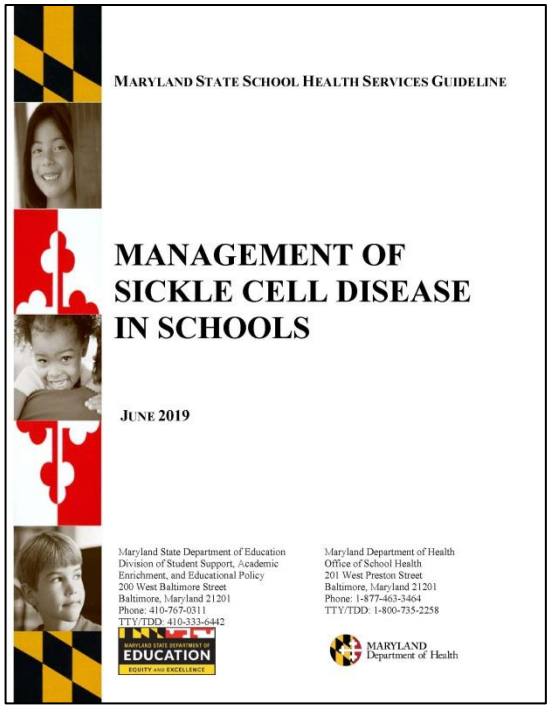
# Role of school personnel

- Communication is key between the school nurse, educators, coaches, bus drivers and other related school personnel that will have contact with a student that has sickle cell disease.
- Understanding the emergency care plan is essential.
- Communication with the student.

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# Responding in the school setting

*Pain*

Students reporting pain should be taken seriously and the school nurse should be notified. The school nurse should have a treatment plan that will include a healthcare provider management plan.

- Rest
- Hydrate
- Adjust temperature conditions
- Receive medication
- Use coping strategies
- Notify caregiver

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## *Severe headache*

A stroke can occur if sickle cells block blood flow to an area of the brain. The nurse should perform the FAST test.


Stroke association

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## *Acute Chest Syndrome*

Acute chest syndrome (ACS) is one serious health complication caused by the blockage of blood flow to the lungs. Symptoms can include fever, cough, chest pain, labored or rapid breathing and shortness of breath. If a student previously had acute chest syndrome or pneumonia, he or she is more prone to have it again.

- Act quickly
- Call 911
- Call parent
- Follow treatment plan
- If student has asthma, follow their action plan for asthma also during this time.

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## *Infection*

Local infections can quickly become systemic because the spleen may be enlarged and there can be scarring.

- Notify the school nurse
- Follow the plan of care that the healthcare provider (hematologist) has ordered
- Administer medication as ordered
- Call parent

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## *Bone disease*

Avascular necrosis can occur due to the lack of blood flow to the joints and cause severe pain. This usually can occur in adolescence and young adulthood.

- Limit the student's mobility, consider classroom location and distance between classes
- Allow for rest periods
- Limit carrying a backpack
- If this is a new symptom, the teacher should send the student to the health room for further evaluation

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## *Eye disease*

Retinal detachment or sudden loss of vision can occur due to deficiency in the amount of oxygen reaching the retina and disease of retinal vessels.

- If this occurs in the classroom, send to health room.
- Notify parent or call 911.

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## *Priapism*

Sickle cells can get stuck in the penis causing a painful, unwanted erection. About 30 percent of males under the age of 20 have experienced priapism. Priapism can cause impotence, so it is important to treat as quickly as possible.

- Give pain medication
- Heating pad
- Drink fluids
- Call parents if lasting longer than 30 minutes

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## *Other potential health problems*

- People with SCD often have a decreased number of red blood cells, a condition called anemia, which can cause lack of energy, breathlessness, and pale color of the skin and lips.
- Sleepiness
- Decreased energy level
- Children reach puberty usually two years later than their peers
- Jaundice and/or gallbladder issues

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# Educational considerations

- 504 or an IEP should be implemented based on evaluations and assessments.
- Each student should have an individualized health care plan (IHCP) as well as an Emergency Treatment Plan.

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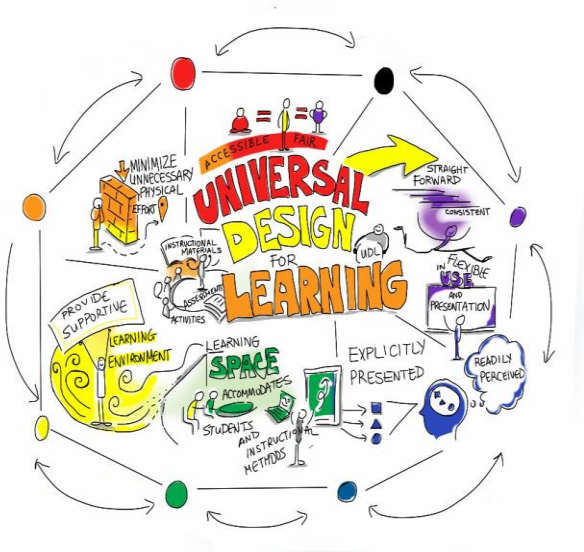
# Accommodations

*Why does a student with SCD need accommodations?*

- Students with SCD have increased absences due to specialist appointments , pain, and other complications or monthly transfusions
- Learning difficulties related to strokes
- Silent strokes - there are no outward signs such as slurred speech or paralysis but there are symptoms of trouble concentrating, forgetfulness, drowsiness, or irritability that can show up.

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## *Plan for extreme temperatures*

- Allow to dress in layers
- Consider alternatives for outdoor recess (above/below certain temperature range)
- Possible blanket for fire drills in cold weather
- Climate controlled environment
- Transportation including pick up, drop off location
- Consideration for classroom temperatures or preferential seating locations (consider windows, vents, fans and heaters).

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## *Avoid dehydration*

- Access to water at all times
- Unlimited bathroom breaks

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*Prevent fatigue and pain*

- Frequent breaks especially during physical education
- Classroom locations
- Elevator key if needed for two story buildings
- Extra set of books at home
- Copies of notes
- Rest breaks or designate a rest area
- Students reporting pain or headache, need to go to school health room immediately.

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## *Student self-esteem*

- Many students with a chronic illness can feel isolated, or that they are the only one with the illness.
- Identify school personnel that the student is comfortable going to if the student has concerns.
- Consider lunch bunches, suggestions of clubs in the school, and support from guidance counselor and school nurse.
- Have a plan if student is out of school due to complications so that they feel connected to their home school.

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# Summary

- School personnel should take any report of pain, fatigue , shortness of breath, and headache from students with SCD seriously.
- It is also important, just like any other student with chronic health conditions, that they be treated as normally as possible.
- With appropriate accommodations, students who have SCD can be successful.

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# Role of the school nurse

*Where to start?*

- Complete a health appraisal/assessment, including baseline vitals, pulse ox, capillary refill time.
- Current treatment plan from Hematologist for emergency care.
- When to contact caregivers, 911 services, and preferred hospital for treatment needs.
- Education of appropriate school staff for emergency actions.
- The school nurse needs to be a part of the 504/IEP team.
- Student specific treatment plan, including pain scale with parameters.
- Medical releases to speak with physicians.

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# Resources

Maryland State School Health Services Guideline- Management of SCD in Schools
http://marylandpublicschools.org/about/Documents/DSFSS/SSSP/SHS/SHSGuidelines/SickleCellDiseaseGuidelines.pdf

CDC- Tips for Supporting the Student with SCD
https://www.cdc.gov/ncbddd/sicklecell/documents/tipsheet_supporting_students_with_scd.pdf

NJ Department of Health- SCD Information for School Personnel
https://www.nj.gov/health/fhs/nbs/documents/Sickle%20Cell%20School%20Personnel%20Guide_Fourth%20Edition.pdf

Virginia Sickle Cell Awareness Program- SCD Handbook for School Personnel
http://scinfo.org/wp-content/uploads/2015/07/SchoolHandbook_SickleCellChild_PDF1.pdf

Specialized Health Needs Interagency Collaboration- Information for School Nurses
https://www.kennedykrieger.org/community/initiatives/specialized-health-needs-interagency-collaboration-shnic/information-school-nurses

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