

# Tips for Supporting Students with Sickle Cell Disease



## Why We Created this Booklet

The purpose of this booklet is to describe sickle cell disease (SCD) and identify roles for teachers, other school staff and parents/caregivers to support students living with SCD. We identify ways SCD may impact a student's daily life and describe how school staff can make accommodations (i.e., adjustments to the classroom setting or instruction) to meet the needs of children who may experience health problems associated with SCD during the school day. The content covers information on ways SCD might impact education outcomes and information about pain, stroke, and symptoms associated with SCD that school personnel should know. Each student's experience with SCD is different and the tips discussed should not replace any medical advice a person with SCD receives from their doctor.

## You Play An Important Role

School personnel and caregivers play an important role in a student's health and academic success. For students living with a chronic health condition like SCD, communication between parents and school officials is essential in supporting positive academic outcomes. As a result, we have developed this customizable booklet. We encourage teachers, students, and caregivers to read all sections and tailor the information that relates specifically to their situation.

## The booklet is organized in the following sections:

**Section 1: About Sickle Cell Disease**

**Section 2: Tips for Teachers, Nurses, and School Administrators**

**Section 3: Tips for Parents and Other Caregivers**

*The findings and conclusions in this report are those of the authors and do not necessarily represent the official position of the Centers for Disease Control and Prevention.*

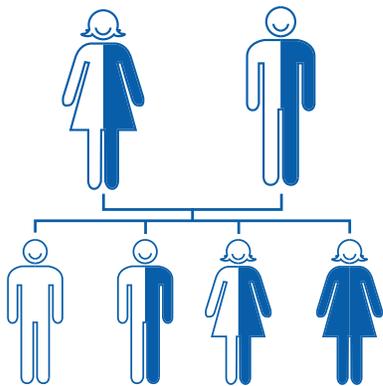
# SECTION 1

## About Sickle Cell Disease

### What is Sickle Cell Disease?

Sickle cell disease (SCD) is an inherited blood disorder (a blood disorder that runs in families). People with SCD produce an abnormal type of hemoglobin (called hemoglobin S (HbS) or sickle hemoglobin). Hemoglobin is a protein in red blood cells that carries oxygen from the lungs to the organs and tissues in the body. The abnormal hemoglobin in SCD can cause the red blood cells to have a sickle or banana shape under certain conditions. 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.

There are many forms of SCD and the most common type is Hb SS, known as sickle cell anemia, which is inherited when a child receives two "S" genes (one from each parent). Hb SC is a form of disease that is inherited when a child receives one sickle cell gene, "S" from one parent and from the other parent, a gene for an abnormal hemoglobin called "C". Another type of SCD, sickle beta-thalassemia, occurs when a child inherits one sickle cell gene and one gene for beta thalassemia (another type of abnormal inherited hemoglobin that causes anemia).



### Sickle Cell Disease in the United States

In the United States, SCD is most commonly found among African-Americans or persons of African descent; however, people of all races and ethnicities can have SCD. About 1 in every 365 African-American babies in the United States is born with SCD; and worldwide, approximately 300,000 babies are born with SCD each year. As more people move from areas highly affected by SCD to the United States, schools will become more diverse and there is a higher chance that teachers will encounter a student with SCD in their classrooms. As with any student with a chronic health condition, students with SCD may experience health problems during the school day.

### How does sickle cell disease affect people?

While normal red blood cells are round like donuts and move freely through blood vessels, sickled blood cells clog the flow of blood and can break apart as they move through blood vessels. Additionally, sickled red blood cells do not deliver oxygen throughout the body as well as normal red blood cells do. As a result people living with sickle cell disease may suffer with:

- Severe pain
- Low number of red blood cells (or anemia)
- Stroke

### What conditions can cause severe pain (sickle cell disease crisis)?

One of the biggest challenges posed by SCD is the unpredictable nature of pain and the wide-ranging severity of health problems due to the condition. Some people with SCD may have infrequent problems with pain, whereas others experiencing pain may require hospitalization.

Certain factors are more likely to trigger a painful sickle cell crisis:

- Infections
- Cold and/or damp conditions
- Air pollution
- Dehydration
- Extreme physical activity
- Stress
- Sudden changes in temperature
- Use of alcohol or caffeine
- Smoking

## SECTION 2

## Supporting Students with Sickle Cell Disease: Tips for Teachers, Nurses, and School Administrators

### What can be done to support students with sickle cell disease?

Teachers, Nurses and Administrators Can:

- 1. Ensure adequate access to water/hydration.** Staying well hydrated by drinking plenty of water can help prevent pain episodes and other health problems. Thus, unlimited access to water throughout the school day is essential. Frequent, small amounts of water are better than trying to drink a large amount of fluid at one time. Allowing access to a bottle of water in class is an option.
- 2. Allow frequent bathroom breaks.** Children with SCD produce large amounts of dilute urine even when they are dehydrated. Thus, children with SCD may need to go to the bathroom more often than other children. Do not restrict students with SCD from bathroom breaks. Provide a special bathroom pass to limit disruptions in instruction and to minimize attention drawn to the student exiting the classroom.
- 3. Allow accommodations during extreme temperatures and conditions.** Cold or hot weather can trigger pain crises. Teachers should not assign a student with SCD a seat in drafty locations, directly in front of fans or under air conditioner vents. Permit layered clothing in the classroom. Remind students with SCD to wear a jacket outside during cold or rainy weather or to take off a layer of clothing when it is hot. Children with SCD should not exercise in extreme conditions (e.g., avoid cold and high heat and humidity).
- 4. Allow accommodations during physical education and recess activities.** Most children with SCD can engage in moderate exercise, including running, swimming, jumping, and riding bikes. However, teachers may modify curricula so that a child experiencing health problems related to SCD can participate in physical education in roles that are less strenuous, such as being the physical education teacher's "assistant," "scorekeeper," or "umpire." Admitting fatigue, which may be due to anemia, may be embarrassing or draw unwanted attention to a child with SCD. Even with moderate activity, regular breaks or a brief period of rest after physical activity may be necessary. In addition, incorporate frequent water breaks into any physical activity plans. Teachers and administrators may want to consult with the child's parent or ask the child the level of activity they can tolerate during recess. Also, remember, never require children with SCD to exercise in cold weather without extra layers of clothing to keep warm.
- 5. Take special care of injuries.** *Never apply a cold pack to an injury or pain site if a child with SCD is injured during the school day.* However, other first aid measures are safe for children with SCD who are injured at school. First aid measures that should be provided when necessary, include applying direct pressure for bleeding, wrapping with an ace bandage, or elevating a hurt limb.
- 6. Watch for signs of stroke.** Some children living with SCD may have learning difficulties due to health problems associated with stroke (blockage of blood vessels in the brain that then causes brain damage). Strokes may be difficult to detect when they affect a small portion of the brain, but they are extremely important to watch for because they are relatively common in the early school years among children with sickle cell disease. Teachers should be aware that declines in academic achievement, inability to maintain attention, difficulties with organization, and mild delays in vocabulary development may be due to small brain injuries caused by strokes. Moreover, teachers are in a unique position to notice changes in school performance that might indicate a stroke and should not simply assume that poor attention in the classroom is due to a lack of the child's motivation or desire to do well in school. Teachers should contact parents when changes in learning or a child's attentiveness are detected so that the child's doctor can be notified. Formal neurocognitive and educational testing may be necessary to determine any learning difficulties caused by stroke. The testing may help school personnel in developing the best teaching strategies for the student. Many students with SCD may qualify for a 504 plan or individualized education plan (see section 3, #2 for more about 504 or IEP). For more information, see the pull out box on stroke.
- 7. Be aware of emotional well-being.** Not all children with SCD have outward signs of illness. However, children with SCD may be smaller in size, have delayed puberty or experience jaundice (yellowing of the skin and eyes). These sometimes subtle, outward signs may make children living with SCD targets for teasing and bullying. Students with SCD may cope with their differences by being aggressive, isolating themselves, or avoiding social situations with peers. Like other children with medical challenges, children with SCD may not have as many opportunities to play with other children, thus recreational activities or group-based

classroom assignments may serve as opportunities for developing good interpersonal skills and boosting a student's self-esteem. Teachers can also help children identify special interests and talents that may help them identify career goals.

#### 8. **Maintain open communication with parents.**

Teachers can help create a positive relationship between home and school as well as a sense of continuity for students by maintaining open communication with a child's family through notes, e-mail, phone contact or conferences to discuss the student's performance and well-being in the classroom and at home. Regular contact with a student's parents is especially important for children with a chronic illness, like SCD. Some children with SCD will have periods

when they are unable to attend school, but may not be hospitalized. Whether a student is hospitalized or homebound due to health problems related to SCD, teachers must allow students the opportunity to complete all the required work. Thus, it may be especially important for teachers to talk to parents about missed school plans prior to a period of illness (tutoring, assignment plan, a second set of books to keep at home or online resources for classwork). When devising make-up work for any child with a chronic medical condition, teachers may want to consider the quality of the assignment over the quantity. Finally, teachers and school nurses should keep up-to-date contact numbers for the student's parents and doctor in case of emergency.

## PAIN EPISODES

Pain episodes are a common health problem of SCD. Pain may occur anywhere in the body (most commonly in the arms, legs, abdomen and back) and may last a few hours, days, weeks or longer. Pain may ease or get worse over this time or may become so severe that a child needs to be hospitalized. The unpredictable nature of pain associated with SCD may create a sense of uncertainty that may cause unusual behavior. A child who is typically easy-going and attentive in the classroom may become grumpy, uncooperative and unresponsive. Many children with SCD suffer with pain on a daily basis and still behave "normal," focusing on their school work and engaging in classroom activities. It is important to recognize when a student's pain is worse than usual or no longer tolerable.

Best practices: **Be responsive to complaints of pain.** A student will come to know whether their pain is mild or moderate and will pass, or whether it is necessary to call his or her parents or go to the hospital. If a child has pain during the school day, schools can help by allowing the child time to rest and then returning him or her to class, if possible. Do not encourage the student to simply bear the pain. In clinic or hospital settings, doctors often use a scale of 1-10 to rate pain. Use this type of scale to check in periodically with a student to learn the difference between the student's usual level of pain and when emergency contacts need to be called.

**Create an individualized care plan for each student with SCD.** Care plans should have input from the teacher, school nurse, the student, and his or her family. Plans may include instructions about giving pain medicine, including who is responsible for giving the medicine, and how to decide which medications to give to the student. It is important to refer to the rules of your local school district when developing plans regarding giving medicines to students during the school day. Students with SCD are often prescribed strong medications, including narcotics (e.g., Tylenol with Codeine, Darvocet), anti-inflammatory medications (e.g., Toradol, Advil) or steroids (e.g., Prednisone) to relieve pain. It is not always in the student's best interest to leave school or be absent due to pain; however, *always notify parents if their child's health status changes during the school day.*

## FEVER

Children with SCD are at greater risk for certain bacterial infections compared to other children. A fever of 101° Fahrenheit (38° Celsius) or higher, could mean the child has an infection. Infection is the leading cause of death in young children with SCD, and frequently leads to hospitalization. If fever is accompanied by pain in the ribs or chest, coughing, and difficulty breathing, this may be a sign of acute chest syndrome, a serious medical emergency.

Best practice: **Be alert for signs of fever.** Teachers should have access to an individualized care plan for a student with SCD that lays out instructions for what to do if a fever occurs during the school day. Children with SCD and fever should be seen by a doctor to evaluate the child for health problems, such as pneumonia or other infection; so contact the student's parent(s) immediately.

# STROKE

SCD is one of the most common causes of childhood stroke. Strokes are small brain injuries that may impact learning. Between 10% and 20% of children with SCD will have a symptomatic stroke, meaning that the stroke will produce physical changes in the student. Most commonly, a stroke can happen if sickled red blood cells get stuck to walls of blood vessels and clog blood flow to the brain. Signs of stroke include severe headaches, dizziness, visual changes, sudden onset of weakness (not due to pain) in one limb or side of the face, numbness, typically in the face or the limbs, sudden inability to produce speech, or a seizure. More than 25% of children with SCD suffer from a silent stroke by six years of age. Silent strokes do not have the same clear signs of symptomatic strokes, but may be evidenced by changes in a student's behavior, concentration, or sudden decline in the quality of their schoolwork. Silent strokes are a serious problem that may require consultation with a doctor who specializes in the brain and behavior.

Best practice: **Be aware of signs of stroke.** Teachers are in a unique position to identify changes in students' behavior that may be linked to symptomatic or silent strokes associated with SCD. If a sudden stroke is suspected, parents should be contacted immediately and the child should be rushed to the hospital. Children who have suffered a stroke or whose medical tests indicate that they are at higher risk of stroke may need monthly hospital appointments to receive blood transfusions to prevent strokes. Create plans to help children make up missed school work. If a student with SCD begins to show gradual declines in academic performance, attention, or memory, teachers should contact the student's parents so that medical attention can be sought if necessary.

## When to seek medical care for students with Sickle Cell Disease

Sudden or worsening symptoms, like chest or abdominal pain, fever (>101 degrees), or any sign of stroke (e.g., weakness or numbness on either side of the body, not able to talk, sudden dizziness or headache, difficulty with memory, blurred vision) require immediate medical help. Remember, always notify parents if their child's health status changes during the school day.

### Signs and Symptoms of Sickle Cell Disease

Sign/Symptom	Description	Call 911
<b>Pain or discomfort</b>	Headache (severe)	✓
	Chest pain	✓
	Bone/joint/ hip pain	
	Upper left, abdominal pain (spleen)	✓
	Priapism (sustained, unwanted erection)	✓
<b>Paleness</b>	Noticeable change in the color of the skin, lips, fingernails	
<b>Fever</b>	101 degrees or higher (indicates a medical emergency)	✓
<b>Weakness or fatigue</b>	Weakness on either side of body (may be due to stroke)	✓
	Inability to speak (may be due to stroke)	✓
	Difficulty with memory (may be due to stroke)	
	Sudden or constant dizziness (may be due to stroke)	✓
	Blurred vision	✓
	Vomiting	
<b>Changes in Breathing</b>	Difficulty breathing	✓
	Fast rate of breathing	✓
	Harsh noisy breathing	✓
<b>Swelling</b>	Hands	
	Feet	
*NEVER APPLY ICE*	Joints (with redness)	

✓ parents should be contacted immediately and 911 may need to be notified

## SECTION 3

### Supporting Students with Sickle Cell Disease: Tips for Parents and other Caregivers

#### What can be done to support students with sickle cell disease?

Parents and Caregivers Can:

- 1. Set up a meeting to discuss SCD with your child's teacher.** It may be important for parents and teachers to meet at the beginning of each school year. Invite these school staff to an introductory meeting: your child's teacher(s) including the physical education teacher, learning coordinator, school nurse, principal, or counselor. During an introductory meeting, it may be helpful to describe what SCD is in broad terms (e.g., an inherited disorder that affects the amount of oxygen carried in the bloodstream) and more specifically how SCD impacts your child's daily life. It may also be important to describe how your child copes with having SCD and to discuss clearly how you wish to be informed when your child has symptoms at school (e.g., call me immediately, when to call paramedics, what hospital you prefer).
- 2. Talk to teachers about a 504 plan or Individualized Education Plan (IEP).** A 504 plan requires that a child with special healthcare needs (i.e., that limits one or more major life functions) be provided with ways to participate in general education programs. For example, in a 504 plan, students and families may develop a written request for access to an extra set of books for home, more frequent bathroom breaks, access to water throughout the school day, or the need for extra layers of clothing. A 504 plan is administered in schools that receive federal funds (i.e., public schools) and is a written document that outlines reasonable accommodations for individuals with disabilities. The 504 plan ensures that a child with SCD has equal access and is able to participate fully in school activities; however it does not outline plans for remedial instruction. The IEP addresses remedial instruction. When health problems related to SCD negatively impact a student's academic performance, special education services may be recommended. An IEP is a written document developed between school staff and families to assure specialized or remedial instruction. Additional federal funding is given to schools to provide a wide range of services, depending on the needs of the child. Both an IEP and 504 plan should be updated yearly to meet the student's needs.
- 3. Develop an individualized care plan.** An individualized care plan is a written document that can be tailored to the specific healthcare needs of a student with SCD. An individualized care plan requires



input from the teacher, school nurse, the child, and the family. Plans should include emergency contact information, any special needs the student may have, and instructions for giving pain medication, including who is responsible for giving the medicine and how to decide which medication to give. Review the school district's policy statement about school staff giving medications. Update care plans with the school nurse at least yearly, or whenever your child's healthcare needs change. If the school does not have a nurse on staff, often the school secretary or counselor may be responsible for coordinating an individualized care plan.

- 4. Tell teachers about changes in your child's health.** Families can support their child's school success by keeping in close contact with teachers. Parents should talk to teachers about plans in the event their child misses school (tutoring, assignment plan, a second set of books to keep at home or online resources for classwork) before a period of illness occurs. These plans can be spelled out formally within a 504 plan. When your child is sick, parents can inform the school, ask for lesson plans and homework, or ask for homebound teachers to prevent the student from falling behind in their coursework. A homebound instructor may be provided by your school district to help students make up schoolwork missed during occasional and extended absences due to hospitalization or health problems that make it difficult to attend school.

Even more importantly, parents should speak with their child's teacher when the student is feeling well, to develop a relationship when things are calm. Keeping in contact when a student is not ill helps teachers to tell the difference between a student's typical behavior, and performance that might signal health, learning or behavioral problems. In addition to an introductory meeting at the start of the school year, parents and teachers should check-in with one another as the school year progresses, through phone calls or in-person meetings, to talk about the child's health status. Finally, parents may also want to talk to teachers about whether or not it's okay for them to tell anyone about their child's condition, or explain why their child is absent to classmates or other teachers, in light of their child's right to privacy. As children grow, their ability to express their needs evolves. In the early elementary school years, parents may discuss all of their child's healthcare needs with teachers. As children get older it is important to support them in becoming their own advocate. Parents can help their child understand their condition, help them to understand the ways that the school is required to help them keep up with their coursework and reduce the risk of health problems related to their condition, and they can encourage their child to express his or her medical needs clearly.

### **What are some common questions that parents or caregivers might be asked by school staff?**

Parents and caregivers may be faced with questions from school staff regarding sickle cell disease. Here are some commonly asked questions and answers that might help parents when having conversations with school staff:

**Q: *Is sickle cell disease contagious?***

**A:** No, you cannot catch sickle cell disease like a cold. Sickle cell disease is a genetically inherited disorder, passed down from a person's parents.

**Q: *Why is your child out of school so often?***

**A:** A person with sickle cell disease needs to be seen by a doctor more frequently than other students, so they may be at a doctor's appointment. At other times, sickle cell disease may cause a person to be in so much pain that he or she cannot attend school.

**Q: *Why do people with sickle cell disease have yellow eyes?***

**A:** Sickle cell causes a person's red blood cells to die more quickly than the red blood cells of a person who does not have sickle cell. Their eyes become yellow due to a substance that is released when the red blood cells break down.



**Q: *Why should a student with sickle cell disease be able to keep a water bottle at his or her desk or leave class more frequently for water fountain and restroom breaks?***

**A:** Water helps to increase a person's vein size and allows sickle-shaped cells to flow through blood vessels more easily. Allowing a student with sickle cell disease to access water freely may help to reduce the pain that can occur with sickle cell disease. As a result of needing to drink so much water, and because sickle cell disease causes kidney problems, the student may need to use the restroom more often.

**Q: *Why are children with sickle cell disease smaller or less physically developed than other children?***

**A:** Children with sickle cell disease have red blood cells that do not carry oxygen as well as children with normal red blood cells. In order for any child's body to grow and develop, oxygen is needed throughout the body for energy. A child with sickle cell disease will grow and develop at a slower pace because less oxygen is being delivered throughout his or her body by the red blood cells.

## Helpful Resources

Adams RJ, Ohene-Frempong K, Wang W (2001). Sickle cell and the brain. *Hematology Am Soc Hematol Educ Program* 2001 (1): 31–46. doi:10.1182/asheducation-2001.1.31

Dyson S. (2011). Sickle Cell and Thalassemia Health and Safety: A Guide to School Policy. Retrieved on March 1, 2013 from <http://www.sicklecellanaemia.org/teaching-resources/resources/scooter24-29/scooter25.html>.

New England Pediatric Sickle Cell Consortium. Sickle Cell Disease: A Resource for the School Nurse and Educator. Retrieved on March 1, 2013 from [http://www.nepscc.org/NewFiles/teacher\\_and\\_school\\_nurse\\_12\\_04.pdf](http://www.nepscc.org/NewFiles/teacher_and_school_nurse_12_04.pdf).

Schatz J, Roberts C, Robinson J, McClellan C. (2004) Sickle Cell Disease: Information for Parents, Teachers, and Schools. Retrieved on March 1, 2013 from <http://www.kagoon.com/sickle-cell-disease-information-for-parents-teachers-and-schools/main>.

Virginia Department of Health. Understanding the Child with Sickle Cell Disease: A Handbook for School Personnel. Retrieved on March 1, 2013 from [http://www.vdh.virginia.gov/ofhs/childandfamily/childhealth/cshcn/sickleCell/documents/2012/pdf/publications/SchoolHandbook\\_SickleCellChild\\_PDF\[1\].pdf](http://www.vdh.virginia.gov/ofhs/childandfamily/childhealth/cshcn/sickleCell/documents/2012/pdf/publications/SchoolHandbook_SickleCellChild_PDF[1].pdf).

Link to Centers for Disease Control and Prevention (CDC) information on Sickle Cell Disease <http://www.cdc.gov/ncbddd/sicklecell/facts.html>.

Link to the U.S. Department of Education (DOE) information on IEP <http://ed.gov/parents/needs/speced/iepguide/index.html>.

Link to the U.S. Department of Education (DOE) information on 504 Plans <http://www2.ed.gov/about/offices/list/ocr/504faq.html>.