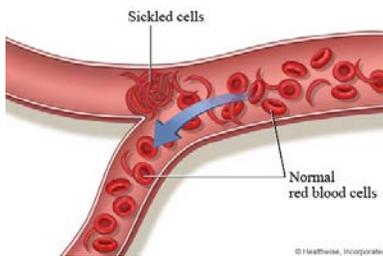


Sickle Cell Disease *At a Glance*

Sickle cell disease (SCD) is an inherited blood condition that is characterized by pain crises and anemia. In the US, newborn screening in most states can diagnose sickle cell disease, usually before the onset of symptoms. The diagnosis of SCD is made by testing for the presence of significant quantities of an abnormal hemoglobin, known as **Hemoglobin S (Hb S)**.

People with sickle cell disease have abnormal hemoglobin molecules (Hemoglobin S). They cause 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 (curved), they break down prematurely leading to anemia.



Normal red blood cells are smooth and can change their shape to flow through 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

About one in every 5,000 babies is born with sickle cell disease, mostly affecting people of African descent. In the US, about 1 out of 500 African American children born will have sickle cell disease. It is important to realize, however, that sickle cell disease affects individuals of different races and ethnic backgrounds.

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.

Things to Think About

1. Medical / Dietary Needs

What you need to know

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.

Up to date immunizations are very important. A yearly check up and studies as needed should occur in the child's Medical Home. Yearly vision screening can be done at the school. The school should contact the children's primary care doctor before allowing them to join in sports.

No special diet is required for sickle cell disease, although a well balanced diet is important. If a child with sickle cell disease (SCD) is eating poorly it is important to let parents know. Hydration is very important. Children with sickle cell disease should be allowed unrestricted access to drinking water. Caffeine causes blood vessels to restrict so should be avoided.

NOTE: It is important to be sensitive to cultural differences in diet.

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. If blood flow is reduced in even a small area of the body, 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.

Pain can be anywhere and can vary in length, location, intensity, and time between episodes.

- Pain medication and hydration is important for treatment.
- Pain crises are the most frequent cause of sickle cell disease-related hospital admissions as well as school and work absences

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Anemia

Anemia is caused by the altered structure of the red blood cells. This change causes the cells to break down more rapidly, leading to 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 (yellowing of eyes and skin).

Dactylitis (swollen fingers and toes)

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 restriction in the blood supply (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. This occurs in 10-30% of children with SCD. Children with spleen issues may experience abdominal pain, nausea, and vomiting. Signs of spleen problems include: irritability, unusual sleepiness, appearing pale, weakness, rapid heartbeat, big spleen, pain on the left side of the abdomen.

Priapism

This is painful and unwanted erections which often start in childhood and often occur in the early morning hours.

Infection

Some infections that may occur in individuals with SCD are:

- Septicemia (infection of the blood)
- Meningitis (infection around the brain) with pneumococcal and other bacteria
- Pneumonia (infection in the lungs)
- Osteomyelitis (infection of the bone)
- The most common cause of death in children with SCD is Streptococcus pneumonia sepsis. Death risk highest in the first few years of life.

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- 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 (similar to pneumonia) is caused by red blood cells trapped in the lungs. This is a form of acute lung injury in patients with SCD and a major cause of mortality. It can develop rapidly. 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 is caused by an infection caused by Parovirus B19. This infection causes red blood cell (RBC) production to be shut down for 10 days. The RBC in children with SCD only live for 10-15 days compared to normal red blood cells that live 120 days. Therefore with this infection, the blood count drops. 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.

Bedwetting

Bedwetting may occur from age 3-adulthood in some individuals. It is more common in boys.

What you can do

Children who have SCD should have access to the nurse's office as requested.

Avoidance of climate extremes is important. 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.

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.

2. Education Supports

What you need to know

It is important to have HIGH EXPECTATIONS for learning for children who have Sickle Cell Disease.

Having sickle cell disease alone does not affect learning ability, but the complications of the disease may cause learning problems. Students may require extra help and encouragement when illness interferes with school.

Fatigue due to anemia and sleep disturbances may be present. This can cause memory problems and a lack of concentration. This can limit a child's ability to focus and pay attention in a classroom setting.

What you can do

Consider an IEP or 504 plan. If a child has learning difficulties, developmental and/or neurocognitive assessments may be appropriate. Some children have attention or other learning problems that appear to be related to stroke, severe anemia, or silent cerebral infarction. Frontal brain injury in SCD is the most common location of injury and can lead to:

- Reduced efficiency in rehearsing verbal information in working memory
- Difficulties with manipulating verbal information in working memory
- Poorer retrieval of verbal information on memory recall trials
- Difficulties with focusing and sustaining attention
- Keeping organized

Keeping up school work is important. Plan for absences so that the child can have work to do away from school. Consider tutoring. Monitor work so that it is challenging, but there are attainable and realistic goals. Communication with parents is important to meet these challenges.

Communication with parents about any change in the child's abilities is important. There may be a need for additional evaluations by the pediatrician or other specialist for any new brain injury.

Individuals with sickle cell disease can participate in school activities and in athletics. However, because of the issues of anemia and fatigue and their sensitivity to dehydration and to cold temperatures, care must be taken to insure their safety.

- Avoid overheating.
- Easy access to water, bathroom, and rest should be available at all times.
- During physical activity, students with sickle cell disease should be encouraged to drink extra amounts of water and other fluids.
- Because anemia is part of the disease, they may fatigue more quickly than other students and may need extra time to recover from activity.

3. Behavior & Sensory Support

What you need to know

Individuals with SCD are small for age and have delayed sexual maturation. Puberty in individuals with SCD tends to occur later and last longer. This can affect self-esteem and body image. It is important to reassure individuals that although it may take longer they will complete puberty and that most adults with SCD are of normal height.

Individuals may be perceived as younger due to their size.

What you can do

Stress management support may be helpful. A guidance counselor and/or psychologist may be supportive to child and/or family. Children who have SCD may also benefit from support for developing positive self-esteem.

4. Physical Activity, Trips, Events

What you need to know

Individuals with sickle cell anemia can participate in school activities and in athletics. However, because of the issues of anemia and fatigue and their sensitivity to dehydration and to cold temperatures, care must be taken to insure their safety.

Sickle cell carriers:

Although sickle cell carriers or those with what is called sickle cell trait are not a focus of this educational tool, you should be aware of the controversy in the sickle cell community about those individuals and their ability to compete in athletics.

- In April of 2010, primarily as a result of legal actions related to a death of a college football player, the NCAA adopted a policy requiring Division I institutions to perform sickle cell trait testing for all incoming student athletes.
- This policy has been met with opposition in the sickle cell community as well as within the medical community, due to the lack of clear scientific evidence or studies and the risk of stigmatization.
- Both the American Society of Hematologists and the Sickle Cell Disease Association of America have come out with statements against testing as a prerequisite for participation in athletic activities and recommendations for implementation of universal interventions to reduce exertion-related injuries and deaths.

Links to learn more about this topic:

- **National Collegiate Athletic Association (NCAA) Sickle Cell Trait resources and FAQs**
<http://www.ncaa.org/health-and-safety/medical-conditions/sickle-cell-trait>
- **National Athletic Trainers' Association** - "Sickle Cell Trait and the Athlete" Consensus Statement
<http://www.nata.org/NR062107>
- **Sickle Cell Disease Association of America (SCDAA)** – “Sickle Cell Trait and Athletics”
<http://www.sicklecelldisease.org/index.cfm?page=sickle-cell-trait-athletics>
- **American Society of Hematology** - “Statement on Screening for Sickle Cell Trait and Athletic Participation”
<http://www.hematology.org/Advocacy/Statements/2650.aspx>

What you can do

- Emphasize hydration and make sure that there is access to water and other fluids for all activities
- Avoid extremes of temperature (i.e., swimming in cold water or playing in snow can trigger a pain episode).
- Build up the exercise program slowly and encourage conditioning programs

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- Avoid performance tests such as mile runs, or serial sprints
- Allow the individual with sickle cell disease to set their own pace
- Stop the activity if muscle cramping, pain, swelling, inability to “catch their breath” or fatigue occurs

5. School Absences and Fatigue

What you need to know

Children with sickle cell disease may have frequent hospitalizations and doctor’s appointments. Pain can cause a child with sickle cell disease to miss school, even without a hospitalization. Anemia may cause a child to tire more easily. Easy fatigability will occur more in the heat.

What you can do

Make sure the child is well hydrated and can have a place to rest when they become fatigued. Students may require extra help and encouragement when illness interferes with school. Keeping up school work is important. Monitor work so that it is challenging, but there are attainable and realistic goals. Plan for absences and consider tutoring. Communication with parents is important to meet these challenges.

6. Emergency Planning

What you need to know

The child needs 24 hour access to a medical facility that can provide urgent evaluation and treatment for acute illness such as fever, acute chest syndrome, and splenic sequestration (over-filled spleen).

Learn more about 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. The medical term for this is splenic sequestration. This occurs in 10-30% of children with SCD.

Children with spleen problems may experience abdominal pain, nausea, and vomiting.

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Signs of spleen problems include:

- Irritability
- Unusual sleepiness
- Appearing pale
- Weakness
- Rapid heartbeat
- Big spleen
- Pain on the left side of the abdomen

What you can do

A plan is needed to deal with mild to moderate episodes of pain. Application of heat, hydration and/or ibuprofen may provide relief. A plan should be in place if fever, chest pain, respiratory symptoms, and/or behavior changes are seen. Be aware if a child has a severe headache or is groggy. It could be a sign of a stroke. Contact parents and hospital as necessary.

7. Resources

Sickle Cell Disease Association of America (SCDAA)

<http://www.sicklecelldisease.org/>

This organization has envisioned a national coordinated approach to addressing things related to sickle cell disease since 1971. There are member organizations across the country.

Genetics Home Reference

<http://ghr.nlm.nih.gov/condition/sickle-cell-disease>

Consumer-friendly information about human genetics from the U.S. National Library of Medicine

Sickle Cell Information Center

<http://scinfo.org/>

The mission of this site is to provide sickle cell patient and professional education, news, research updates and world wide sickle cell resources.

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Kid's Health from Nemours - Sickle Cell Disease

http://kidshealth.org/parent/medical/heart/sickle_cell_anemia.html

KidsHealth is the #1 most-visited website for children's health and development.

Sickle Cell Kids

<http://www.sicklecellkids.org/>

This site is for kids with sickle cell disease to have a way to connect with others.

GeneReviews

<http://www.ncbi.nlm.nih.gov/books/NBK1377/>

Learn more about the genetics of Sickle Cell Disease.

American Academy of Pediatrics (AAP) – Health Supervision Guidelines

<http://pediatrics.aappublications.org/content/109/3/526>

The AAP endorses these Health Supervision Guidelines. Families may find these helpful when talking to their pediatricians or family physicians

Centers for Disease Control and Prevention

[Tips for Supporting Students with Sickle Cell Disease](#) was created by the CDC in order to provide a tool for teachers, other school staff, and parents/caregivers to support students living with SCD.

Note: *This printable version does not include the information found under the green button marked “Transitions” on the website. Those general pages may be printed separately.*

Meet Amarey, a Child with Sickle Cell Disease

GEMSS would like to thank Amarey and her mother for their generosity in sharing this story with us. You have made the site come to life with the addition of your thoughts and feelings. Thank you so much!

Amarey is a smart, vibrant little girl who, at 3 ½ years old, is a ball of energy and a whiz with technology. She can find Nick, Jr. on a lap top computer, type in the codes to get into her mom's iPhone, seeks out YouTube videos on a Kindle, and is eager to teach other children how to play games on her Leap Frog! "She is so happy" says her mother, Amarilis. "She loves to dance, sing and memorize songs."



Amarey will be heading to preschool in her local school system in the fall, her first school experience. After interviewing many preschools in her New England city and discussing Amarey with her school administrators, Amarilis has chosen a school that fits Amarey's particular needs. The school has a nurse and the school is air-conditioned all year round to help keep her body cool. In choosing a school, it was also important that the school was very clean and staff were vigilant about sanitizing toys and play areas. Her mother feels like she is ready to let go of her fear of Amarey getting sick and let Amarey experience the joys and opportunities offered at school. In fact, in preparation, Amarey will be heading to day camp at the YMCA for 6 weeks this summer and will be enrolled in dance classes too. Amarilis also thought about people being CPR/first aid/epipen certified as she was visiting schools.

Amarey does not qualify for an IEP but she will have a 504 Plan. She will also have a Medical Action Plan available for the school to help with medical needs and emergencies. Her school plan will allow for Albuterol, Epipen, and Motrin for pain if any of those are needed. Amarilis also expects teachers to be alert for signs that Amarey may be getting ill such as a decrease in her playing and socializing, fatigue, or skin coloration changes that might signify jaundice. It will be important for Amarey to be able to have water with her or nearby for frequent drinks to keep her well-hydrated.

Amarey was diagnosed with Sickle Cell Disease (type SS) within the first week of her life. Although she has been hospitalized, on average, about every 4 months for various infections, her family is grateful that she recently had an 8 month gap without being hospitalized. When she needs to be hospitalized, it usually starts with a fever, and a temperature of 101 degrees will be reason for admittance. Once ill, she might develop Acute Chest Syndrome, or have low

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hemoglobin requiring a blood transfusion, low oxygen or respiratory infections. She also has many allergies and is asthmatic. She takes medications daily at home.

Amarilis says that their family support system is fantastic. She also says that her confidence has grown as she asks more questions, attends conferences, joins in support groups on Facebook, and gets involved in volunteer efforts to help others with Sickle Cell Disease.

Amarilis says that she hopes that other parents will not be afraid to send their child to school. She advised parents to talk to school's early childhood team, the school nurse, and administrators to make sure everyone is aware and educated on SCD. "Keep asking questions," Amarilis says. She has come a long way in growing her own knowledge and confidence and feels that she has to "give Amarey the life she deserves! I feel I was blessed with her!"