

SICKLE CELL DISEASE

A Resource Guide for Patients and Families



NEPSCC

New England Pediatric Sickle Cell Consortium

www.nepsc.org



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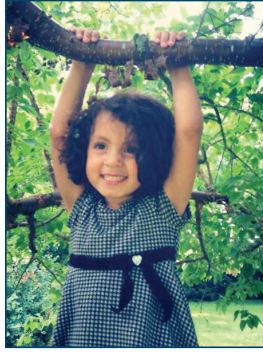
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TABLE OF CONTENTS

INTRODUCTION	Page 1
MEDICAL ISSUES	Page 3
HOW IS SICKLE CELL DISEASE TREATED?.....	Page 8
WHEN YOUR CHILD IS IN PAIN	Page 11
EMOTIONAL ISSUES.....	Page 14
EDUCATION.....	Page 19
ADDITIONAL SUPPORT/RESOURCE LIST	Page 23



INTRODUCTION

Children and families living with sickle cell disease (SCD) often have many questions. This guide was created as a resource to help answer some of those questions and give families the knowledge they need to live and thrive with SCD. *If you have responsibility for someone who has sickle cell disease, it is important that you familiarize yourself with what is in this book before they are in crisis.*

What is Sickle Cell Disease?

SCD is an inherited blood condition that affects red blood cells. (It is not an infectious disease that can be caught.) In people with SCD, red blood cells—which carry oxygen and nutrients throughout the body—are shaped like sickles or crescent moons in stressed conditions (in steady states, many of the RBCs are not sickled) rather than being round and donut-shaped. The sickled red blood cells also become rigid and sticky rather than being flexible and slick losing the ability to move rapidly throughout the body providing needed elements to all organs of the body.

A child will have SCD if he or she inherits two genes that lead to ‘sickling’—one from each parent. The genes involved can be two ‘S’ genes, an ‘S’ and a ‘C’ gene or an ‘S’ and a variant ‘V’ gene. Some people believe SCD originated in West and Central Africa. Some believe it to have originated as a biological evolutionary trait for protection from malaria in locations where the malaria organism was abundant. Whatever the case, SCD has been in existence for thousands of years, perhaps. It is not man-made or contagious. It is a genetic condition. It is caused by the inheritance of two abnormal genes that can only be passed from parents to child/ren. It is not a bacteria, a virus, or another contagious or communicable trait. The carrier gene is widespread and early researchers found it common in locales near the equator. Now, with more widespread research and screening, even, that finding may not be the whole story.

Although, SCD is believed to be a disease largely carried by peoples of African-descent alone, this is not, in fact, true. SCD is a global disease affecting a global population. A 21st century discussion of the disease would wisely note that SCD is found everywhere in the world and that the carrier trait is not carried by persons of

African-descent alone. Screenings need to be undertaken amongst more of the world population, but it has been identified in people who are of African, Asian, Caribbean, Central American, European, Mediterranean, Middle Eastern, North American, Saudi Arabian, South American, and South East Asian descent. Some countries have not reported SCD. Perhaps, in those cases, because screenings are not undertaken. Or, in some cases, because health information is maintained nationally or regionally. However, no one should assume that SCD is not part of their genetic history—the human race’s evolutionary and migratory histories make that assumption highly questionable.

Some time ago, the total number of sickle cell disease sufferers in the United States was estimated at 100,000. Unfortunately, we do not have accurate information for the country as a whole. Only a limited number of states have reasonably accurate counts at present. Each year about two thousand children are born with SCD in the United States.

SCD can affect many organ systems of the body. When sickle-shaped red blood cells get stuck inside smaller blood vessels, this can lead to severe pain at times. Important organs like the brain, heart, and kidneys—which need a constant supply of blood to work properly—can be damaged by sickled cells. Therefore, parents, care-givers, and patients should strive to get care within 30 minutes or less of a sickle cell episode. To do this, it is best to have a care plan in place with your doctor(s) and hospital(s). In the meantime, some doctors are working with hospitals toward creation of *hospital protocols* that ensure patients with SCD are seen as quickly as would an asthma patient. Sickle cell pain can be come ‘status’ (like, status asthmaticus) in the same way that can asthma. Some risks like stroke, organ failure, and death, are too great to take. Awareness assists preparedness. (Contact doctors, hospitals, and advocacy programs to find how your family can be best prepared, treated, and nurtured.) Your child/ren can live, long, happy, healthy and very successful lives with preparation, preventative measures, and appropriate care.

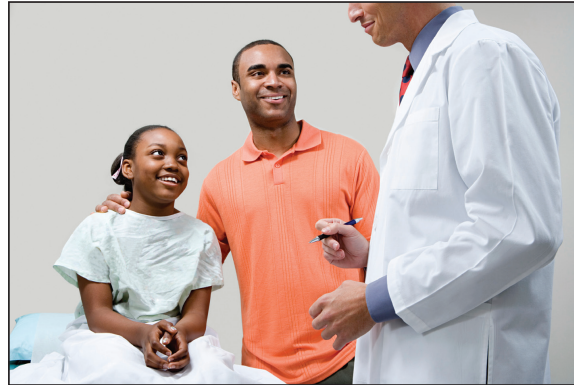
Parents should not be shamed or blamed for passing SCD, in any form, to their child/ren. As in any case, they should strive to be the best parents that they can be (some opt to take courses on healthy well-being for patients suffering chronic disease, some take care-giving classes, some take parenting classes online or in classrooms). But, neither the parents or the children are to be blamed or shamed or negatively labeled for passing or possessing a gene that may provide a protective trait aiding survival.

Genetic and reproductive counseling are comparatively new options and, often, not advertised. There is no reason for your child/ren to live anything other than a lively, productive, successful life—as long as they are provided good care, nutrition, medication, positive environs, and, most of all, information about their disease.

At the appropriate age, male and female adolescents or young adults should undergo genetic and reproductive counseling to address the chance of them passing the gene to their future child/ren, particularly, in the painful “SS” form. In this way, parents

and patients can make informed decisions independently and/or in consultation with a competent and informed medical provider. This, relatively new, approach is becoming more commonplace for many people, of any background, seeking to have children.

Healthy families lead to a healthy society. Let's discuss.



MEDICAL ISSUES

SCD can lead to many medical issues that may vary depending on a child's age and condition. The information that follows addresses some of these medical complications and steps you can take to ensure safe and proper care for your child.

If you notice your child is experiencing the following medical conditions, please contact your doctor:

- **Fever:** a temperature greater than 100.4 degrees F (38 degrees C) for a baby under 3 months of age, or greater than 101.4 degrees F (38.5 degrees C) for children older than 3 months of age.
- **Pain:** either SCD pain or other pain that has not responded to home pain medicines or is different in nature from usual sickle cell pain.
- **Breathing:** any new breathing problem: cough, wheezing, shortness of breath, tightness of the chest or chest pain.
- **Fussiness:** for a baby unexplained fussiness or change in feeding pattern.
- **Vomiting or Diarrhea:** more than one episode in a day or more than one day of vomiting and/or diarrhea.
- **Abdominal Distension:** an increase in abdominal / spleen size.
- **Color:** an increase in paleness or yellowness of the skin or the color of the lips or white of the eyes.

- **Movement/Activity:** any change in strength, ability to use arms or legs, abnormal uncontrolled movements (seizures) or change in consciousness, even if very short lasting.
- **Priapism:** an erection of the penis that persists more than 40-60 minutes or is painful. It is important that you and your child/ren do not allow this event to be sexualized.

Remember that people with SCD may become sick very quickly. Never hesitate to call your doctor if you have any concerns about a possible complication of your child's SCD.

Fever

Fever is a serious risk in sickle cell patients. If your child suffers from SCD, you should make sure that you always have a thermometer handy so you can take his temperature.

Fevers can be caused by viruses, bacteria, and other conditions. Penicillin or another antibiotic is usually prescribed for SCD patients under the age of 5 to reduce the risk of bacterial infection. Taking antibiotics, however, is not a guarantee against bacterial infections. It is also strongly recommended that they be vaccinated against pneumococcal infections. The vaccines used are called Pneumovax® and Prevnar®. While very useful, these too are not 100% guaranteed to prevent such diseases.

If your child develops a fever (a temperature greater than 100.4 degrees F for a baby under 3 months of age, or greater than 101.4 degrees F for children older than 3 months of age), call the doctor immediately. Do not administer Tylenol®, do not wait for the fever to “break,” and do not wait to see if the temperature comes down. If the fever does not persist, call anyway. The doctor may recommend that you take your child directly to the emergency room for treatment. If you go directly to the ER, it can be helpful to notify your doctor ahead of time that you're going.

Pain

Your child should have an individualized pain plan which your doctor can draw up in consultation with you and having evaluated your child. SCD can cause such severe pain that only opioid medication can effectively reduce it. Note that prescriptions for such strong pain medication as hydrocodone, oxycodone and morphine cannot be called in; you must get the physical prescription from the doctor and present it at the pharmacy for it to be filled.

If your child has a pain episode, follow his or her pain plan. This should include drinking extra fluids, using warm baths or hot packs, and doing relaxation exercises. Pain medication should also be used as needed. If the pain does not improve, contact the doctor. It may be that the pain can best be managed in a hospital setting – ER, inpatient or outpatient. You will need to work with the doctor to determine if the

pain responds well enough after one or two pain treatments in the ER or clinic, or if your child should be admitted for pain management.

Pain medications will likely cause mood changes. In fact, the entire SCD episode can evoke mood changes because of the limited flow of blood and the changes in blood chemistry. Parents should expect mood changes and not view these as inherently “bad” characteristics of the child. The child is not naturally “bad”. The child’s physical being is undergoing physical challenges at the time of an SCD episode as well as the period before and after. The child should be encouraged toward self-discipline so that these experiences do not cause emotional spirals or (self-) blame, shame, or labels. Mood changes are an unfortunate side-effect of variations in cells and blood flow to organs. Therefore, the child’s mood may fluctuate in a way beyond the child’s intent or control.

Chest Problems

Sometimes SCD related problems in the chest might make your child very sick, very quickly. If your child develops any new chest problems, such as coughing, wheezing, tightness of the chest, shortness of breath or chest pain, please call to the doctor right away. Your child may need to be evaluated with a chest X-ray and labs or to be admitted to the hospital to receive antibiotics and medical care. If your child has asthma or a history of wheezing, make sure you use the asthma medications as prescribed. Do not try to treat new chest problems at home on your own. Chest problems may indicate a more severe concern. This may indicate “acute chest syndrome” and this event has a higher morbidity rate.

Unexplained Fussiness/Change in Feeding/Vomiting (in babies)

Babies cannot tell you if they are not well or are in pain. It is important, therefore, to be able to recognize warning signs that may indicate your baby is not well. Not feeding as usual, crying more than usual or not sleeping as usual may be a sign of medical problem. If you notice these signs you should call your doctor for advice.

In a younger child, swelling of the hands or feet may be caused by dactylitis. This is when sickle cells become “stuck” in the small blood vessels of the hands or feet. This is often painful and you may need advice on how to make your baby more comfortable.

If your baby is fussy, irritable, not eating as usual, has puffy hands and/or feet or you feel an enlarged spleen, please call your doctor right away.

Increase in Spleen Size

Young people with SCD sometimes develop an enlarged spleen. The spleen is normally a smallish organ situated on the posterior part of left side of the body, inside the lower ribcage. It is usually too small to feel, but if it becomes enlarged, it can become much larger and can be felt easily below the bottom left rib. While it is normally full of blood, in this condition it contains much more blood than usual. In SCD, this is called splenic sequestration, and it is a serious condition.

The doctor should show you where your child's spleen sits, how to feel for it, how to recognize if it is enlarged, and whether to seek medical attention. If you think you can feel your child's spleen, call the doctor right away.

Splenic sequestration does not always make the spleen large and palpable. Sometimes increased fussiness or irritability can be the main clue. Please be on the lookout for these clues and if you suspect a problem, call the doctor.

Increase in Skin Paleness

The phrase "sickle cell anemia" is well known. Not all SCD involves anemia, but it is a common component of the disease. Patients with strong anemia ("lack of blood") tend to become pale. This anemia can result from spleen problems or a viral infection which can cause bone marrow to stop producing red blood cells. This is called an "aplastic crisis."

Paleness is not always easy to spot. Fatigue, headaches, dizziness and shortness of breath can also be signs of an aplastic crisis, as can fussiness or irritability and splenic enlargement.

If your child is pale or shows other signs of anemia, call the doctor right away.

Neurological Changes

Children with SCD have an increased risk of problems with the blood vessels of their head. Because sickle cells can get stuck in the vessels of the brain, people with SCD are at an increased risk of having a stroke, or a transient ischemic event (a short-lasting brain event). To help to identify children at risk, children with more severe forms of SCD are recommended to undergo an ultrasound of his/her head yearly. If this study is abnormal recommendations will be made on how the risk of stroke might be reduced. Your doctor needs to know about physical change suggestive of a stroke, to be able to take care of your child quickly. If your child experiences any weakness of arms or legs, altered consciousness, difficulty in speaking or thinking, or any seizure-like activity (abnormal, uncontrolled movements), you must call your child's doctor immediately. If your child has lasting altered consciousness, do not take the time to call your doctor, but dial 911 or the emergency number for your area and have your child taken to hospital, where your doctor can then help.

If your child has any neurological changes, including weakness, altered consciousness, difficulty speaking or thinking, or seizures, call your doctor or 911 or the emergency number for your area immediately.

Vomiting or Diarrhea

Children with SCD may become dehydrated if they experience vomiting or diarrhea. The sickle cells may “sickle” more if there is less fluid in the blood vessels.

It is important for good health for individuals with SCD to drink plenty of still fluids such as water or Gatorade® or PowerAde® (not sodas). Vomiting and diarrhea lead to loss of body fluid. If your child does experience any vomiting or diarrhea and can take fluid by mouth, clear liquids such as water, Gatorade®, PowerAde®, punch, iced or hot clear tea, or similar liquids are better than a milk-based product at this time. If your child experiences vomiting or diarrhea, call your child’s doctor for advice. Your child may need to be admitted to hospital for intravenous fluid.

Sustained or Painful Erection of the Penis

This is called priapism and is another complication of SCD. This is a complication that needs to be approached in a very sensitive and understanding manner. Priapism in males of nearly any age will almost certainly raise all kinds of issues about their relationship to their parts most closely associated with that gender identity. It is really important that boys experiencing priapism are not teased (or there is an expression of misguided admiration or jealousy) and as a care provider to a boy experiencing priapism, to make sure that the boy is not bullied. It is important that boys with SCD know that this is an extreme of normal that they can experience because they have SCD, that if long lasting (more than 40-60 minutes) it needs to be reported to his doctor.

If a boy does have a painful or prolonged erection of the penis, please be sure to contact the doctor right away for advice. Sometime just pain medicines and fluid will help resolve this problem. Other times, however, a blood transfusion or surgery is necessary. If this problem is not addressed quickly, and particularly if the problem recurs, it is possible that impotence would be the eventual outcome.



HOW IS SICKLE CELL DISEASE TREATED?

The goals of managing SCD are to PREVENT pain, infections, and other complications of the disease. There is no single best treatment for all people with SCD. Treatment options are different for each person depending on the symptoms, but can include medications to treat infections, help with pain, and help make red blood cells. Other therapies include blood transfusions.

A person with SCD can live a full life and enjoy most of the activities that other people do. The following tips will help you stay as healthy as possible:

Prevent Infections — Common illnesses, like influenza, quickly can become dangerous for a person with SCD. The best defense is to take simple steps like washing your hands frequently to help prevent infections and getting a flu vaccine every year.

Learn Healthy Habits — Drinking 8 to 10 glasses of water every day and eating healthy food will help to maintain hydration and proper nutrition. People with SCD should maintain a balanced body temperature, getting neither too hot nor too cold. Participating in physical activity to help stay healthy is very important. Also beware of hiking, mountain climbing or places in high altitudes as they carry less oxygen.

Nutrition and Sickle Cell Disease

Research shows that a person with sickle cell disease requires more calories and nutrients than people without sickle cell disease. Many Americans, with or without SCD, do not meet minimal requirements for good nutrition. For children and adults with SCD, attention to healthy eating is even more important. It is recommended that you focus on nutrient dense foods--these are high vitamin foods and can help make the most of every meal and snack. Specifically, choose foods rich in protein, fiber, and healthy fats such as fruits, vegetables, whole grains, nut butters, and olive

oil. Plenty of fruits and vegetables provide necessary vitamins and minerals such as calcium, folate, and zinc as well as protein and fiber. Finally, drink plenty of water to maintain hydration and prevent complications related to SCD.

Feeding children presents its own unique challenges. Limiting sweeteners, in particular, is really important. Seek out substitutions that will be attractive and fulfilling. For example, sweeteners such as natural and minimally refined sugars like stevia, honey, or maple syrup are excellent substitutes; however, parents must be aware that even these sugars should be used sparingly. It's also important to limit "empty calories" such as those found in chips, other processed snack foods, and sugary beverages. These foods do not provide any essential nutrients necessary for good health, and can cause your child to feel full, making them less likely to eat the nutrient dense foods they need to promote wellness. Nutrient rich juices and smoothies can be used as part of a well-balanced diet, and can help meet those high nutritional requirements of people with SCD. They are also fast and efficient and easy to consume. Remember, a parent can add dark leafy vegetables and fresh or frozen fruit as well as nutritional supplement powders to these juices. (Consult your doctor or medical nutritionist or dietician about your child's best options and alternatives.) Any dietary changes should be considered in consultation with an individual's medical team.

Penicillin — Penicillin helps prevent infections. Children with SCD should take penicillin (or another antibiotic prescribed by a doctor) every day until they are at least 5 years of age.

Vaccines — Vaccines are a great way to prevent many serious infections. Adults and children with SCD should have the influenza vaccine every year, as well as vaccines against pneumococcus and meningococcus—bacteria that cause pneumonia and meningitis-- as well as any others recommended by their doctor.

Folic Acid — Some doctors may prescribe a vitamin called folic acid. It is a vitamin that sometimes makes red cells stronger.

Hydroxyurea — hydroxyurea is a medicine that has been shown to help adults and children with SCD. Studies have shown that hydroxyurea can decrease the number of pain events, pneumonia, acute chest syndrome, and admissions to the hospital, while increasing energy, height and weight, and lifespan. The U.S. Food & Drug Administration approved it in 1998 for treatment of Sickle Cell Anemia in adults, although it has been used since the 1960s for other diseases. Hydroxyurea has been used in children with SCD for over 20 years, with no severe side effects seen in children ranging in age from 6 months to 15 years. Hydroxyurea comes as a capsule or liquid, and is taken by mouth once a day. It must be prescribed by a doctor and is fully covered by most private drug insurance policies and state insurances. You can get your hydroxyurea directly from the pharmacy.

In patients with SCD abnormal sickle hemoglobin causes the red blood cells to become long, rigid “sickle shaped” and sticky. Sickling causes blockage of blood flow to vital organs, muscles, and tissues. With hydroxyurea, red cells have more fetal hemoglobin, become larger, less sticky, and travel more easily through blood vessels. Hydroxyurea also decreases the number of white blood cells, which causes less inflammation and sickling.

People with SCD may be eligible for hydroxyurea treatment starting at nine months of age. Even those with milder disease may benefit since the treatment may prevent sickle complications.

Transfusion — Transfusions have been found to be beneficial in certain clinical situations in order to lessen complications associated with SCD. Use has been most effective in the prevention of initial and recurrent stroke in patients with SCD who are at high risk for this complication. Donor red blood cells contain normal hemoglobin (HbA), which allows the cell to maintain a round and flexible donut shape. Transfusion of donor cells into patients with SCD will decrease the percentage of circulating abnormal sickle cells resulting in improved blood flow and delivery of oxygen throughout the body.

Since not every condition will benefit from transfusion the decision to utilize transfusion therapy in a patient with SCD should be discussed with your hematologist and based on risk: benefit assessment.

Bone Marrow (Stem Cell) Transplant — Stem Cell Transplant is the only curative therapy available for SCD at present. Hydroxyurea and chronic red blood cell transfusions may lessen the complications of SCD, but they do not cure the disease. Over last 4 decades Stem Cell Transplant has been used for the treatment and cure of variety of cancers, blood disorders, immune deficiency disease and certain inherited metabolic disorders. Results of many studies show that transplants from matched related donors offer about an 85 percent chance of cure. As per American Society of Hematology, Stem Cell Transplant is considered a standard of care for the patient with SCD who has a matched sibling donor. Almost all insurance companies cover the cost of transplant. Those without a matched sibling donor may have the option of an unrelated donor transplant or a haplo-identical transplant from their parents or siblings. Transplant evaluation is recommended for all SCD children and adults who experience an aggressive course (recurrent acute chest syndromes, strokes, end organ complication or multiple pain crisis). Please ask to see a transplant physician or a team member, to get more details about stem cell transplant for SCD: <https://bethematch.org/for-patients-and-families/learning-about-your-disease/sickle-cell-disease/>



WHEN YOUR CHILD IS IN PAIN

Pain can be a life-long challenge for those living with SCD. In addition to medical intervention, there are a number of non-medical approaches that may help your child better cope with pain.

To best help your child when he/she experiences pain, you should take responsibility for managing the pain. You should be alert to when your child starts having pain and, at that time ask him/her to rest, to stay warm, to take a warm shower or use a heating pad. You should give Tylenol® or Motrin® as your doctor will have advised. It is often helpful if your child drinks clear liquids at the time of a painful crisis. If the pain does not settle with Tylenol® or Motrin® and you have stronger medicines at home, use the medicines as your child's doctor has prescribed. If your child's pain continues even after you have given your child the prescribed pain medicines, call your child's doctor. It may be that your child's pain would be best managed in the hospital.

While you are trying to resolve your child's pain, you may want to try one or more of the following techniques. Some of these you may be able to provide yourself. Others you will need to ask your doctor about.

Acupuncture is based on the belief that life forces move through the body in specific paths. These paths are called meridians. With acupuncture, a needle is put into the meridian that runs to the area where your child is having pain. This needle blocks the meridian, which stops or decreases the pain.

Aromatherapy is a way of using good smells to help your child relax and decrease pain. Candles, massage oil, scented bubble baths, and even baking cookies are all ways that smells are used. Scientists are learning that good smells may change your child's mood and help him relax. It may also help your child's brain make special chemicals like endorphins, which can help decrease pain.

Breathing exercises are another physical way to help your child's body relax. Teaching the body to relax helps lessen pain. You should teach your child to breathe in and out very slowly. A fun way to practice breathing slowly is to blow soap bubbles or a party blower. Your child will know he is doing great when he gets large bubbles or the blower makes long noises.

Control often helps children have less pain when they need to have medical procedures. If they understand what is going to happen and are allowed to help, they may experience less pain. For example, if possible let your child choose which arm is used for a blood sample. Or, let him put on the bandage. Also, caregivers may explain treatments and procedures using a doll to help decrease the fear of the unknown.

Distraction teaches children to focus their attention on something other than pain. Watching TV, playing board and video games, or telling stories may relax you and your child. This can help keep your child from thinking about the pain. Take a "fun bag" with these kinds of toys when your child goes for treatments or procedures.

Guided imagery teaches your child to put pictures in his/her mind to help make the pain less intense. With guided imagery, your child learns how to change the way her body senses and responds to pain. Ask your child to imagine floating in the clouds, or remembering a favorite place, or imagine doing a favorite activity. Ask your child to tell you what she feels when floating, if she can see things all around, and so on.

Heat can help decrease pain. Some types of pain improve best using heat. A warm bath may help calm your child and let his muscles relax. A heating pad held against the area of pain may help to reduce the pain.

Music It does not matter whether you and your child listen to it, sing, hum or play an instrument. Music increases blood flow to the brain and helps your child take in more air. Scientists are learning that music increases energy and helps change your child's mood. Music also may cause your child's brain to make special chemicals like endorphins.

Relaxation and biofeedback teach your child's body to respond in a different way to the stress of being in pain. Normally, when pain starts, the body responds with tense muscles, a faster heartbeat, and higher blood pressure. Your child's breathing also becomes faster and shallower. These responses can make the pain worse. Relaxation helps make the pain less by changing these responses. Caregivers may use a biofeedback machine so that you and your child know right away when your child's body is relaxed.

Massage is often used to help a child become more relaxed. You or other family members can gently massage your child's back, shoulders, and neck. Massage may work even better if you help your child use guided imagery, breathing exercises, or music.

Being in a quiet place may make it easier for your child to deal with the pain. Avoiding bright lights or loud noisy places may help you control your child's pain. Wrapping

your child up in a blanket and rocking him/her can help him relax. Offering pacifiers and comfortable positioning (if possible) may also help relax your child and decrease pain.

Self-hypnosis is a way for older children to change their level of awareness. This means that by focusing their attention, children can move away from their pain by making themselves open to suggestions like ignoring the pain or seeing the pain in a positive way.

Hypnosis can give long-lasting relief of pain without affecting a child's normal activities. Self-hypnosis gives your child better control of his or her body. Your child may feel less hopeless and helpless because he or she is doing something to decrease the pain.

Please remember that your child may react to how you are feeling If you are upset or nervous, your child may become upset or nervous. This can increase your child's pain. Relaxation exercises can work to help you become calm. If you can be calm and relaxed, your child may become calm and less fearful, which will help decrease his or her pain.





EMOTIONAL ISSUES

Helping Your Child Cope

A chronic illness like SCD can create many challenges for children and their families. As a parent or caregiver you may be asking:

What does my child know about sickle cell disease?

How does my child feel about the treatment?

How can I support my child?

Children of different ages understand and react differently to SCD and its treatment. Your child's personality, normal coping style, support system, treatment plan, as well as age or developmental level will affect how your child copes with SCD.

When children are faced with stress their normal behaviors may change. They often become more dependent on adults or may act younger than their age (baby talk, wetting pants after being potty trained, sucking his/her thumb or holding a favorite toy or blanket.). Your child may not know how to handle the many feelings that have been caused by the SCD diagnosis.

You are a very important part of your child's life for many different reasons. You know what your child has experienced in the past and how your child usually handles stress. You can help the health care team understand your child. You and the health care team can work together to find new ways to help your child cope with SCD and treatment.

In the following section, we look at common ways children of different ages respond to stress and SCD. The section also discusses specific suggestions for how to help children in each age group cope.

INFANT (birth – 12 months)

Infants look to their parent(s)/caregivers to meet their needs. They rely on adults for food, comfort, play, and care. Infants learn about the world around them through

their senses (i.e. can sense new smells, colors, tastes) and trust in people and things that are familiar. Infants have no concept of the meaning of SCD or its implications. They do respond to the new people and the environment around them.

Tips for supporting your infant when he/she is ill or hospitalized

- Leave a shirt with your smell on it if you need to go
- Keep security objects, like blankets, pacifiers, or a favorite toy nearby
- Let your child explore toys with hands and mouth (check for small pieces that could cause choking)
- Use gentle touch and massage to comfort your infant
- Talk to and play games like peek-a-boo with your infant as you would at home
- When in the hospital, ask that procedures be done in the treatment room
- Wake your infant before a painful procedure
- Continue or develop familiar feeding, bedtime, and bath time routines, like rocking, touch, and singing

TODDLER (12 months – 3 years)

Toddlers are beginning to want to do more on their own. Your toddler's favorite words may be "me do" or "no." Growing toddlers need to be able to do some things by themselves to promote a sense of control. Toddlers show you how they feel through their actions because they do not have the words to describe their feelings. They have a hard time understanding how the body works. Toddlers tend to think that they make things happen. They can create their own false ideas about how they got sick and what happens to them ("I hurt because I was bad").

Tips for supporting your toddler when he/she is ill or hospitalized:

- When you leave, tell your toddler where you are going and when you will be back
- Provide security objects like a blanket or stuffed animal
- Let your toddler make choices whenever you can (ex: apple juice or orange juice?); however, do not offer a choice when no choice exists
- Give your toddler a job to do (ex: holding a band aid)
- Try to keep eating, sleeping, and bathing routines as normal as possible
- Give your toddler safe ways to express anger and other feelings (ex: Play-Doh, painting, building blocks)
- Set limits with your toddler and discipline when needed
- Keep security objects, like blankets, pacifiers, or a favorite toy nearby
- Use simple words, pictures, or books to tell your toddler what will happen before a treatment or procedure

PRESCHOOLER (3-5 years)

Preschoolers are also trying to do things on their own. They take pride in being able to do things for themselves (“I can do it!”). Preschoolers are learning more words to express what they think and feel. However, they often use their play to tell you these same things. They can see the hospital and treatment as punishment for something they did wrong. Also, they often become confused by adult words and make up reasons for the things that happen.

Tips for supporting your preschooler when he/she is ill or hospitalized:

- Tell your child what will happen a little before the treatment using simple words, pictures, or books
- Explain terms that your child may not understand (ex: thinking a “CAT” Scan has a cat or looks for a cat)
- Let your child play with doctor kits and safe medical supplies such as a blood pressure cuff
- Allow your preschooler to make choices whenever you can (ex: apple juice or orange juice?); however, do not offer choices when choices do not exist
- Praise your child for doing things independently such as dressing, brushing teeth, and eating
- Maintain your routines as much as possible as a child responds best to predictability
- Use play to help your child to show feelings
- Maintain discipline strategies as much as possible as before diagnosis

SCHOOL AGE (6-12 years)

School age children take pride in being able to do most things by themselves. They enjoy school because it helps them learn and master new things. Their friends are becoming more important. School age children are able to think in terms of cause and effect and have a better sense of time. They have more words to describe their bodies, thoughts, and feelings. School age children can also understand more of how their bodies work; however, they still may have a hard time with and be confused by medical words.

Tips for supporting your school-age child when he/she is ill or hospitalized:

- Allow your school-age child to make choices whenever you can, but do not offer choices when no choices exist
- Let your child practice things that are new and scary
- Let your child go to school or do school work and activities
- Maintain the same routines and discipline for the whole family
- Have your school-aged child write letters, e-mail or call friends
- Let friends visit when your child feels well enough

- Use simple works, pictures, or books to tell your child what will happen
- Let your child play with safe medical supplies like a blood pressure cuff

TEENS (13-18 years)

Teens are beginning to see themselves as individuals in the world. They are striving to be independent from the adults around them. As teens strive to think and act for themselves, their peers become even more important. Teens want to be like their friends and are concerned with how they are viewed by others. Illness and treatment cause teens to be different when they are trying so hard to be the same. Teens are able to see not only cause and effect, but also can see things from many points of view.

It is important for you to help your child feel secure, protected, loved, and safe as the hospital can seem or feel foreign. Over time, as your child grows, assisting your child to navigate hospital environs independently will be important and help the transition from pediatric to adult care, from childhood to maturity. Take it in steps. It is a developmental process. Your doctor as well as the *transition teams* in pediatric and adult hospital programs can help. In time, your child should feel defined by who they are and what they have overcome to feel like complete, independent, capable, inherently stable, encouraged adults despite the challenges of SCD.

Tips for supporting your teen when he/she is ill or hospitalized:

- Let your teen be active in social and school activities
- Involve your teen in the treatment plan by including him or her when talking to the team about the plan
- Allow your teen to do things that makes him/her feel good about him/herself
- Respect that teens may need to do some things by themselves, when possible
- Offer your teen private time
- Encourage time with peers and allow friends to visit or call in the hospital or home
- Answer questions openly and honestly
- Help your teen to plan for the future
- Encourage your teen to keep doing normal things like school
- Give your teen safe ways to express feelings, especially anger, such as physical activity, talking, or writing
- Assure your teen that all feelings are normal, such as guilt, fear, or sadness

Impact of Sickle Cell Disease on Siblings

Brothers and sisters of a child with SCD may have many different feelings and responses. Often they have needs similar to their brother or sister with SCD. They may feel upset, scared, and unsure of what the future holds.

While siblings may feel sad and worried about their brother or sister, they may also feel some resentment or anger. Mom and dad are spending all of their time with or talking about their sibling.

Friends and family send gifts and money. Children, especially young ones, may feel jealous. Siblings may also feel sad and cry easily. Often siblings have problems of their own, such as depression, trouble sleeping, physical complaints, or problems in school.

How to help siblings

The following suggestions may be useful in helping your other children cope with their brother or sister's illness:

- Try to spend time alone with your other children, doing things that are of interest to them.
- Let your other children know they are still loved and important to you.
- Talk with your other children about the diagnosis, treatment, and effect on their brother or sister. What you tell them will depend on their ages and ability to understand. Assure them that SCD is not contagious and they are not responsible for their brother or sister getting SCD.
- Take your other children with you to the hospital to help them feel involved in the care and treatments of their sibling. Taking them to the hospital or clinic may help to decrease their fears and help to keep a feeling of closeness with their brother or sister.
- Ask a friend or relative to stay in your home, rather than send your children elsewhere when the child is hospitalized.
- Allow your children to help with chores at home to help them feel needed and help you too.
- Talk with your children's teachers. Teachers can be supportive of your children and let you know about any school-related problems.
- Ask for help from a member of the health care team, such as the child life specialist, social worker, psychologist, or child psychiatrist.



EDUCATION

When children with SCD reach school age, they may face special challenges due to their illness and lack of understanding of the disease by those in the school setting. There are special laws in place to ensure your child overcomes any barriers to receiving an appropriate education. Some of these laws include:

Section 504 of the Rehabilitation Action of 1973/American's with Disabilities Act (ADA)

Section 504: Public School

All children with medical conditions have a right to the same educational experience as their non-disabled peers. Children with SCD may need accommodation or modifications to their school day so that they can equally access their education. A “504 Plan” is a list of accommodations that a child with a disability requires so that they can be successful in school. Speak to the school nurse, social worker, etc. regarding meeting to develop a “504 Plan”

Section 504: College

Section 504 also applies to post-secondary programs. College students have a legal right to accommodations to their SCD so they can be successful in college. Connect your college’s “504 Office” to begin the process.

Individuals with Disabilities Education Act (IDEA):

IDEA is a law for children who have learning challenges. Each child “identified” under IDEA has a special written plan setting out their services and supports to help them be successful in school. Accommodations related to the SCD would be in the IDEA document – you do not need a separate 504 Plan.

School Attendance:

If your child has frequent absences from school, you may want to include this in the 504 or IDEA plan.

You are your child's greatest advocate. By working closely with your child's teachers and other school staff, as well as your child's health care team, you ensure your child receives an education that will enable him or her to thrive despite the obstacles to learning that SCD sometimes presents.

You may wish to distribute the information below to staff at your child's school to help them better understand your child's SCD.

Discrimination:

The American's with Disabilities Act (ADA) ensures that persons with disabilities are not subject to discrimination simply because of their disability and that they have the same opportunity to participate in activities in the public and private sector. For example, a child with SCD cannot be rejected from a public or private camp simply because they are a child with a disability. This is true for all activities.

Programs and Support for Persons with Disabilities:

There are many state and federal programs that provide support and resources for persons with SCD. The programs have a range of eligibility criteria. Some of those programs include:

- Social Security Disability for Adults and Children

- TANF – Temporary Assistance to Needy Families: Some states have cash assistance programs for parents who cannot work because of their or their child's medical condition

- Medicaid Transportation (Non-Emergency Medical Transportation – NEMT) – Patient on Medicaid may be eligible for transportation to medical appointments. The type of transportation you receive should be based on your specific medical needs.

Utilities:

Some States have laws that protect persons with certain medical conditions from having their Utilities terminated. Talk with your child's social worker if you need assistance with the payment of your Utilities.

Understanding Sickle Cell Disease: Instructions for Teachers

For the most part, children with SCD should be able to actively participate in the regular school setting. There are, however, some ways to make participation in day-to-day activities less likely to trigger complications of the disease. It is important

to keep in mind that for children with SCD, any severe or untreated complication can be life threatening. If you observe any sign or symptom that concerns you, you shouldn't hesitate to get the child into the care of medical professionals or evaluated at a hospital or treatment center.

BASIC CLASSROOM GUIDELINES

Teachers should: anticipate more missed school days. A child with SCD often misses more school days than normal because of episodes of pain, routine medical treatments, and trips to the hospital. It's helpful, therefore, to have a game plan for providing make-up work so that she will be able to continue her studies during the absence whenever possible. Aside from any materials or extra help that you can offer, if you anticipate a prolonged absence or see a child starting to lag behind, you might suggest that her parents arrange for a tutor at home or at the hospital.

Teachers should: allow extra water and bathroom breaks. In busy classrooms, it is not uncommon to limit water and bathroom breaks. For children with SCD, however, these rules need to be relaxed a little. The simple act of staying well hydrated can help prevent pain episodes and avoid organ damage. Children with SCD also need to take bathroom breaks more frequently, not only because of increased water intake but because their kidneys produce more urine, even when they're dehydrated.

Teachers should: pay attention to temperature control. Often becoming overheated or too cold is enough to trigger a pain episode in a child with SCD. Keeping the temperature in the classroom comfortable is important, as is reminding the child to wear a jacket outside during cold weather or to take off a layer of clothing if she gets hot.

Teachers should: be aware of the need to rest. Because of the anemia characteristic of SCD, children can become tired sooner than their peers. For many children, admitting to this fatigue and taking a break from sports and gym activities can be embarrassing and draw unwanted attention. While participation should be encouraged, make it easy (and as inconspicuous as possible) for the child with SCD to take regular breaks.

Teachers should not: assume lesser academic abilities. While SCD can affect many aspects of everyday life, it does not inherently play a role in a child's intelligence. Missed school and the impact of having a lifelong illness, but not generally from a learning disability, may affect the academic abilities of a child with SCD. For this reason, it is especially important to promote continued learning even in the face of absences, and identify learning problems as you would for any other child. In this day and age, children with SCD can lead long and productive lives, and a good education is key to making this a reality.

Teachers should: be vigilant. SCD is one of the few conditions that can cause stroke in children. An uncharacteristic slip in academic performance may be a subtle sign.

Please notify the child's family at once if any of the following symptoms appear without another explanation:

- Difficulty with memory
- Difficulty using a hand, a leg, or one side of the face due to muscle weakness
- Numbness or tingling
- Difficulty with balance
- Difficulty with vision or hearing
- Difficulty speaking or understanding what other people are saying
- Unusual headache

Other possible causes of school problems include adjustment difficulties after absences, side effects of pain medications, depression, family conflicts, and fatigue.

Teachers should: be informed. The best thing you can do is to be informed about SCD and then create a plan to help a child stay involved, free of complications, and engaged in learning. Children with this disease are, in most ways, just like other children, but they do face certain particular challenges because of their lifelong disease. You can play an important role in offering them the chance to lead relatively normal and productive lives.



ADDITIONAL SUPPORT

Sickle Cell Resource List

There are many resources available to help you and your family face life with SCD. You may find the information below helpful.

A diagnosis of SCD may be overwhelming and extremely stressful at times. The ways people respond to the diagnosis and treatment vary, and many social, emotional, and spiritual concerns may arise. A pediatric hematology social worker is available to provide individual and family counseling that will assist you and your child to help deal with concerns in the following areas:

- Your child's feelings about him/herself
- Reactions to the illness
- School issues
- Financial and insurance concerns
- Providing electric and gas company documentation of medical necessity
- Application for a disabled parking permit

We are here to help.

Sickle Cell Disease Association of America

Website: www.sicklecelldisease.org

The Sickle Cell Disease Association of America provides the latest information on the treatment of SCD as well as research and news about the disease.

Sickle Cell Disease: Information for School Personnel

Website: www.state.nj.us/health/fhs/sicklecell

This information is designed for school personnel and explains many issues related to SCD. This is an excellent site to pass on to teachers who may want to know more about a student's condition, and it's helpful for SCD patients and families as well.

EDUCATION

Federation for Children with Special Needs

Website: www.fcsn.org

The Federation for Children with Special Needs provides information, support and assistance to parents of children with disabilities, their professional partners, and their communities.

FINANCIAL ASSISTANCE

Medicaid

Telephone: 1-800-841-2900 (for general information)

Website: www.hcfa.gov/medicaid/medicaid.htm

Medicaid: this is a Federal program of comprehensive medical coverage for low and moderate-income residents. Many kinds of coverage are available. Some programs include prescription drug coverage, as well as transportation assistance to medical appointments.

If you are eligible for Medicaid you most likely are eligible for the following service (please check with your primary care doctor or social worker): Free door-to-door transportation for your child to medical appointments. Your child's physician should complete the application. Ask your physician or social worker how to obtain an application.

Women, Infants and Children Nutritional Support Program (WIC)

This program supports low-income woman and children up to age five years by providing foods to supplement nutritional needs.

The Special Supplemental Nutrition Program for Women, Infants, and Children (WIC) provides Federal grants to States for supplemental foods, health care referrals, and nutrition education for low-income pregnant, breastfeeding, and non-breast-feeding postpartum women, and to infants and children up to age five who are found to be at nutritional risk.

Website: <http://www.fns.usda.gov>

Supplemental Security Income (SSI)

Supplemental Security Income (SSI) is a Federal income supplement program funded by general tax revenues (not Social Security taxes):

It is designed to help aged, blind and disabled people who have little or no income and it provides cash to meet basic needs for food, clothing and shelter.

Children with SCD are eligible for SSI benefits depending on the family income. SSI also makes referrals to other benefit programs.

Website: <https://www.ssa.gov>

NEPSCC

New England Pediatric Sickle Cell Consortium

BAYSTATE MEDICAL CENTER

Springfield, MA

<https://www.baystatehealth.org/services/pediatrics/specialties/hematology-oncology>

BOSTON MEDICAL CENTER

Boston, MA

<http://www.bmc.org/pediatricshematology-oncology.htm>

BOSTON CHILDREN'S HOSPITAL

Boston, MA

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

CONNECTICUT CHILDREN'S MEDICAL CENTER

Hartford, CT

<http://www.connecticutchildrens.org/our-care/cancer-and-blood-disorders/>

FLOATING HOSPITAL FOR CHILDREN

Boston, MA

<https://www.floatinghospital.org/Patient-Care-Services/Departments-and-Services/Hematology-Oncology/Overview.aspx>

HASBRO CHILDREN'S HOSPITAL

Providence, RI

<http://www.hasbrochildrenshospital.org/pediatric-hematology-oncology.html>

BARBARA BUSH CHILDREN'S HOSPITAL AT MAINE MEDICAL CENTER

Portland, ME

<http://www.mmc.org/pediatrics-cancer-care>

MASSACHUSETTS GENERAL HOSPITAL FOR CHILDREN

Boston, MA

<http://www.massgeneral.org/children/services/treatmentprograms.aspx?id=1610>

UMASS MEMORIAL MEDICAL CENTER

Worcester, MA

<https://www.umassmemorialhealthcare.org/umass-memorial-medical-center/services-treatments/childrens-medical-center/z-list-pediatric-services/hematology-blood-diseases>

UNIVERSITY OF VERMONT MEDICAL CENTER

Burlington, VT

<https://www.uvmhealth.org/medcenter/Pages/Departments-and-Programs/Pediatric-Hematology-and-Oncology.aspx>

YALE - NEW HAVEN CHILDREN'S HOSPITAL

New Haven, CT

<https://www.ynhh.org/childrens-hospital/services/cancer-and-hematology.aspx>

WYCKOFF HEIGHTS MEDICAL CENTER

Brooklyn, NY

<https://www.wyckoffhospital.org/departments/clinical /pediatrics>

COMMUNITY ORGANIZATIONS:

CITIZENS FOR QUALITY SICKLE CELL CARE; SICKLE CELL DISEASE ASSOCIATION OF AMERICA, NORTHERN CT

New Britain, CT

www.cqsc.org

GREATER BOSTON SICKLE CELL DISEASE ASSOCIATION, INC.

Boston, MA

www.gbscda.org

SICKLE CELL DISEASE ASSOCIATION OF AMERICA, SOUTHERN CT

New Haven, CT

www.sddaasouthernct.org

The current revision of this booklet was compiled with the help of many of the members of the NEPSCC who deserve many thanks. Individual thanks must go to the following who provided significant help and insight to allow this version to be in the format that hopefully provides a useful tool for our patients, their families and the community dedicated to the care and success of managing SCD:

Biree Andemariam, MD, UCONN Health Center, Comprehensive Sickle Cell Center, Farmington, CT

Natasha Archer, MD Boston Children's Hospital

Laura Goldstein, Psych.D Boston Medical Center

Julio Martinez, MD Boston University School of Medicine

Amelia Lord, Certified Holistic Health Coach

Caitlin Neri, MD, MPH Boston Medical Center

Pat Pisciotto, MD American Red Cross (retired)

Niketa Shah, MD Yale New Haven Children's Hospital

Amy Sobota, MD, MPH Boston Medical Center

Roger Thrall, MD Hospital for Special Care, New Britain, CT

Editor: Philippa Sprinz, MD, MSc Hasbro Children's Hospital, RI

Special thanks must also go to Kimberly Syphrett, without whose unwavering support and belief that we would be able to achieve a revision that would be of value to the our SCD community, the new edition would not have been achieved.



*With special thanks to our anonymous
charitable donors.*



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