

# *Sickle Cell Disease Transition Guidebook*



From cute-wallpaper.com.

*Providing youth education and ensuring the  
continuity of care*

## Contact Information

We are here to help! If you need anything please use this guide for who to call and how to best reach us.

### **Pediatric Hematology Clinic Office** (Call: 617-414-4841)

- Either follow the prompts, or enter 1, 3, 2 to reach a person.
- Call this number if you need to schedule, cancel or reschedule an appointment, or for prescription refill requests.
- Someone here can also help you reach your nurse or doctor during the day.

### **BMC paging operators** (Call: 617-414-5000)

They can page the person you are trying to reach to call your phone.

Ask them to page the pediatric hematologist on call - pager 5731

### **Automated paging system** (Call: 617-638-5795)

Enter the pager number of the person you want to page and then enter your call back number. On call pediatric hematologist - pager 5731

### **Providers**

Sprinz Philippa, MD (pager 2587) [Philippa.Sprinz@bmc.org](mailto:Philippa.Sprinz@bmc.org)

Sobota Amy, MD (pager 3253) [Amy.Sobota@bmc.org](mailto:Amy.Sobota@bmc.org)

McMahon Lillian, MD (pager 0400)

Maria Champigny, LICSW (pager 5143) Social Worker [Maria.Champigny@bmc.org](mailto:Maria.Champigny@bmc.org)

### **Pediatric Hematology Clinic**

Yawkey Building, 5<sup>th</sup> Floor

850 Harrison Ave, Boston MA 02118

### **Adult Hematology Clinic/Sickle Cell Transition Clinic**

Moakley Building, 3<sup>rd</sup> Floor

830 Harrison Ave, Boston, MA 02118

Here is who you should call to help you with some frequent problems:

***Who do I call if.....I am having pain?***

During the day call or page your doctor either through the clinic (call 617-414-4841 and then enter 1, 3,2), or through the paging operators (call 617-414-5000 and ask for them to page the pediatric hematologist) or through the automated paging system by calling 617-638-5795, enter pager 5731 to reach the on-call pediatric hematologist and then enter your call back number. At night or on weekends use one of the paging options only.

***Who do I call if.....I am low on pain medication?***

Call the office at 617-414-4841 and follow the prompts (1,3,1,2) to leave a message. Please leave your name and a call back phone number in addition to what medications you need.

Prescriptions for opioid pain medication (such as oxycodone) must be picked up in person.

***Who do I call if.....my PT1 will expire next month?***

Congratulations for being pro-active! Call your social worker to make sure this does not expire.

***Who do I call if.....I can't make my appointment?***

Call the office at 617-414-4841 and enter 1, 3, 2 to reach a person. Ask them to reschedule your appointment.

***Who do I call if.....if I have a general question about sickle cell disease?***

You can always email your doctors with any non-emergency questions, or if you want more information.

## Booking Appointments with Specialists and Primary Care Physicians

### Pediatrics

Specialties	Contact Information
Dentistry	Yawkey Ambulatory Care Center Call: 617-414-4046
Ophthalmology (Eyes)	Boston: 617-638-8350 Brockton: 508-588-3060 Middleboro: 508-947-8868 Taunton: 508-823-7473
Orthopedic Surgery (Bones)	Shapiro Center, 4 <sup>th</sup> Floor, Suite 4B Call: 617-638-5633
Pediatric Specialties Group	Call 617-414-4841 and follow the prompt
<ul style="list-style-type: none"> <li>• Cardiology (Heart)</li> </ul>	Yawkey Ambulatory Care Center, 5 <sup>th</sup> Floor
<ul style="list-style-type: none"> <li>• Developmental and Behavioral</li> </ul>	Vose Hall, 4 <sup>th</sup> Floor
<ul style="list-style-type: none"> <li>• Endocrinology/Diabetes</li> </ul>	Yawkey Ambulatory Care Center, 5 <sup>th</sup> Floor
<ul style="list-style-type: none"> <li>• Gastroenterology (Stomach)</li> </ul>	Yawkey Ambulatory Care Center, 5 <sup>th</sup> Floor
<ul style="list-style-type: none"> <li>• Hematology/Oncology (Blood)</li> </ul>	Yawkey Ambulatory Care Center, 5 <sup>th</sup> Floor
<ul style="list-style-type: none"> <li>• Neurology (Brain)</li> </ul>	Shapiro Center
<ul style="list-style-type: none"> <li>• Pulmonology (Lungs)</li> </ul>	Yawkey Ambulatory Care Center, 5 <sup>th</sup> Floor
Primary Care	Yawkey Ambulatory Care Center Call: 617-414-5946
Renal Medicine (Kidney)	Shapiro Center, 7 <sup>th</sup> Floor, Suite 7A Call: 617-414-8680
Urology	Shapiro Center, 3 <sup>rd</sup> Floor, Suite 3B Call: 617-638-8485

## Adults

Specialties	Contact Information
<b>Cardiology (Heart)</b>	Cardiovascular Center Preston Family Building, 3 <sup>rd</sup> Floor Call: 617-638-7490
<b>Dentistry</b>	Yawkey Ambulatory Care Center, 5 <sup>th</sup> Floor Call: 617-414-4050
<b>Endocrinology</b>	Nutrition & Weight Management Preston Family Building, 2 <sup>nd</sup> Floor Call: 617-638-7470
<b>Gastroenterology (Stomach)</b>	Moakley Building, 2 <sup>nd</sup> Floor Call: 617-638-6525, option #1
<b>Hematology/Oncology (Blood)</b>	Solomont Center for Hematology/Oncology Moakley Building, 3 <sup>rd</sup> Floor Call: 617-638-6428
<b>Neurology (Brain)</b>	Shapiro Center Call: 617-638-8456
<b>Ophthalmology (Eyes)</b>	Boston: 617-638-8350 Brockton: 508-588-3060 Middleboro: 508-947-8868 Taunton: 508-823-7473
<b>Pulmonology (Lungs)</b>	Shapiro Center, 9 <sup>th</sup> Floor, Suite 9B Call: 617-638-7480
<b>Primary Care/General Internal Medicine</b>	Shapiro Center, 5 <sup>th</sup> and 6 <sup>th</sup> Floors Call: 617-414-5951
<b>Renal Medicine</b>	Shapiro Center, 7 <sup>th</sup> Floor, Suite 7A Call: 617-414-8680
<b>Women's Health Group</b>	Shapiro Center, 5 <sup>th</sup> Floor, Suite 5A Call: 617-638-7428

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## Introduction...

When young adults with sickle cell disease transfer their medical and non-medical care to the adult service, they may find it difficult to understand all that is expected of them. This guidebook provides the necessary information and resources to help you transition into the adult care setting successfully. It will also help prepare you for a new role as an independent adult by exploring available medical, clinical, nutritional, and social services. We care about your health and strongly encourage you to make the best use of these resources to keep you living a healthy life with the disease as you grow older.



..... It's all about you

Some information contained in this guidebook was obtained from:

- Center for Young Women's Health website
- Handbook on Sickle Cell and School: Bridging the Gap
- New England Pediatric Sickle Cell Consortium website
- National Heart Lung and Blood Institute website
- Massachusetts Department of Social Services - Handbook for skills development.

## Facts about Sickle Cell Disease

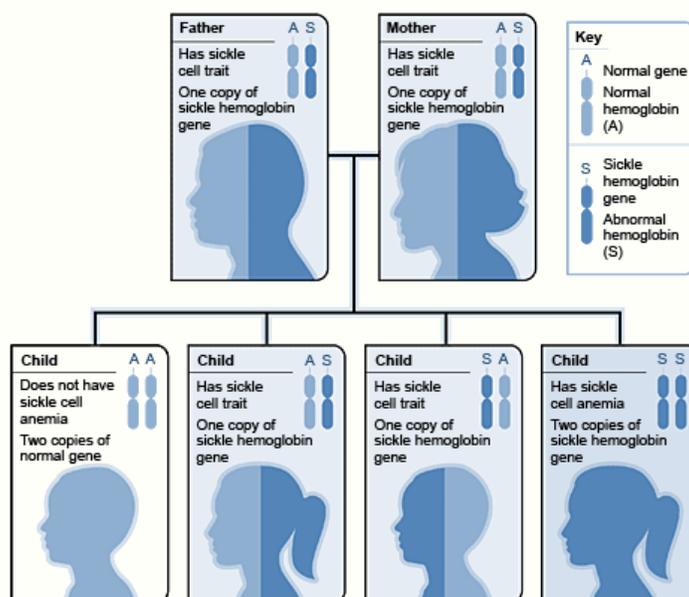
### *How did I get Sickle Cell Disease?*

Sickle cell disease is inherited. That means you inherit one copy of the sickle cell gene from each of your parents.

Everyone has two genes for hemoglobin (the protein in the red blood cells that carries oxygen), one from each parent. “A” is normal hemoglobin, and “S” is sickle hemoglobin.

If both parents have one normal gene and one sickle gene, he or she has “AS”, also known as sickle trait, and their children have a one in four chance of inheriting sickle cell disease (SS). Each child has a 25% chance (1/4) of inheriting two normal genes (AA), a 50% chance (2/4) of inheriting one normal gene and one abnormal gene (AS or sickle cell trait), and a 25% chance (1/4) of inheriting two abnormal genes (SS). Inheriting two abnormal sickle genes gives a child “SS” or sickle cell anemia.

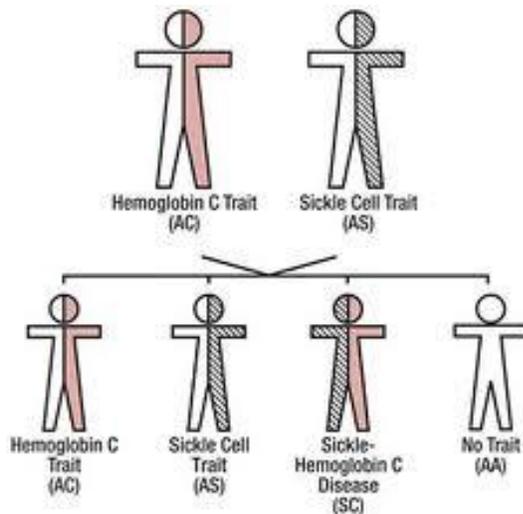
The picture on the next page illustrates how someone can get sickle cell disease (SS) from his/her parents.



From NHLBI <http://www.nhlbi.nih.gov/health/health-topics/topics/sca/causes.html>

Some people have a “C” gene which is another variation of hemoglobin. If you have “SC” then you have a different type of sickle cell disease.

The picture below shows how someone can get sickle cell “SC” disease from his/her parents.



Some people inherit a combination of sickle cell disease and beta thalassemia. Beta thalassemia is a condition which occurs when not enough (beta) hemoglobin is made. If one parent has “AS” (sickle cell trait) and one parent has  $\beta^0$  (Beta) thalassemia trait, there is a 25% chance for each child to have sickle-  $\beta^0$  thalassemia.

### ***Can I pass on sickle cell disease to my children?***

Yes! If you have “SS” and you have children with someone who has sickle cell trait “AS” then each child has a 50% chance (1/2) of having sickle cell disease “SS” as illustrated in the table below.

If your partner has only normal hemoglobin all your children will be carriers of the disease and no one will have the disease.

You can prevent this from being a surprise by making sure your partner knows his or her sickle status. His / her parents may be able to tell your partner, or he/she can ask his/her doctor for a hemoglobin electrophoresis test. If you or your partner were born after the early 1990's in the United States, it is likely that you were screened with newborn screening tests for Sickle Cell Disease and other abnormal hemoglobins. If you were born outside the United States it is possible you were not tested as a baby.

### Sickle Cell Inheritance

If I have...	If my partner has...	Our children may have...
Sickle cell anemia (SS)	Sickle Cell Trait (AS)	50% sickle cell trait (AS) 50% sickle cell anemia (SS)
Sickle cell anemia (SS)	Normal hemoglobin (AA)	100% sickle cell trait (AS)
Sickle cell/hemoglobin C (SC)	Sickle cell trait (AS)	25% sickle cell trait (AS) 25% hemoglobin C trait (AC) 25% sickle cell anemia (SS) 25% sickle cell/hemoglobin C (SC)
Sickle cell/hemoglobin C (SC)	Normal hemoglobin (AA)	50% sickle cell trait (AS) 50% hemoglobin C trait (AC)
Sickle cell/beta thalassemia	Sickle cell trait (AS)	25% sickle cell trait (AS) 25% sickle cell anemia (SS) 25% thalassemia trait 25% Sickle cell/beta thalassemia
Sickle cell/beta thalassemia	Normal hemoglobin (AA)	50% sickle cell trait (AS) 50% thalassemia trait

### ***Can I grow out of it?***

No, your genes do not change. The symptoms of sickle cell disease may change when you get older, but if you are born with “SS” you will always have “SS.”

### ***Is there a cure?***

The only cure for sickle cell disease is a stem cell transplant, usually from a brother or sister with the same mom and dad. Talk with your Doctor to find out if this is an option for you.

### ***What are the complications of Sickle Cell Disease?***

Sickle cell disease affects many organs/systems in the body. Complications are caused by the abnormal ‘sickle’ shape of the red blood cells as well as inflammation and hemolysis (breakdown of red cells). These can often be prevented or minimized if you are well-monitored by your doctor by keeping regular clinic appointments and following up as recommended with specialists. Some of the common complications other than pain include:

- **Acute Chest Syndrome:** Acute chest syndrome is caused by sickled hemoglobin trapped in the small blood vessels of the lungs. This causes shortness of breath, fever and chest pain. It can be triggered by a chest infection such as pneumonia. Acute Chest Syndrome can also develop before, during, or after an episode of sickle cell pain. This is why it is so important to use your incentive spirometer if you are admitted for a pain episode.

Acute chest syndrome is a medical emergency and requires hospitalization and treatment with antibiotics and may require a blood transfusion. If you think you are experiencing acute chest come to the ER immediately.

- **Eye problems:** Sickle cell disease can lead to obstruction of blood vessels in the retina, part of the back of the eye. This can, over time, lead to problems with vision. If this sickle ‘retinopathy’ is detected early it can be treated. For this reason, we recommend that everyone with sickle cell disease have a dilated eye exam by an eye doctor every year.

- **Gallstones:** When the hemoglobin in red blood cells breaks down in the body, it releases bilirubin. In an individual with sickle cell disease, the red blood cells are constantly being broken down and bilirubin forms rapidly. Excessive bilirubin in the body can form hard substances in the gallbladder called gallstones.

People with gallstones usually have pain at the upper right side of the abdomen, under the right shoulder, or between the shoulder blades. They may also have nausea, vomiting, fever, sweating, chills, colored stool or yellowish discoloration of the eyes. If you have repeated episodes of pain from your gallbladder, the Hematologist or the Surgeon may recommend its removal.

- **Infection:** In people with sickle cell disease, the spleen stops working well in childhood, increasing the risk of frequent infection. It is advisable you receive a flu shot every year and get all available vaccines against infections. Always call your doctor or report to the hospital if you have a fever, which might be a sign of serious infection.
- **Leg Ulcers:** People with SCD may have one or more sores on the leg called ulcers. This is often found in the lower third of the legs. Ulcers can sometimes persist for years or re-occur after healing.
- **Priapism:** This is a painful erection that occurs in males with sickle cell disease. It is caused by a blockage of blood flow out of an erect penis. If you experience priapism treat it like you would a pain episode—drink lots of fluids, take a warm shower, use your pain medication, and attempt to urinate. If the erection lasts for more than 1 hour please go to the emergency room since prolonged priapism can lead to impotence.
- **Pulmonary Hypertension:** The blood vessels in the lungs can be damaged by the sickled cells making it difficult for the heart to pump blood through the lungs. This eventually leads to an increased blood pressure in the lungs. This is one reason we may recommend that you see a pulmonary (lung) specialist in your late teen or early twenties, so that this can be detected early.

- **Splenic Sequestration:** The spleen is an organ in the body found in the upper left part of the abdomen, under the rib cage. It helps to separate abnormal red blood cells from the normal ones, and also fights infections. In people with sickle cell disease, the spleen may trap both abnormal and normal red cells. The abnormal cells may block the normal flow of blood out of the spleen.



This makes the spleen become big and sometimes painful. When the spleen traps many cells and an anemia follows this is known as splenic sequestration. Splenic sequestration is more common in young children, but if you have sudden pain in your left upper abdomen call your doctor right away.

- **Stroke:** The blood vessels in the brain can also be blocked by sickled cells. This may reduce the blood supply with oxygen to the brain causing a condition called stroke. Stroke may lead to convulsions, paralysis, lasting brain damage or even death. Common symptoms of stroke include weakness in the arms, feet or legs, slurred speech, changes in vision, severe headache and vomiting. In patients with Sickle Cell Disease a stroke can also be caused by bleeding in the brain. Stroke is common in young children with sickle cell (SS and S beta zero thalassemia) disease and older people with these forms of the disease. When you were younger if you had SS disease or S beta zero disease, your doctor will have ordered Transcranial Doppler (TCD) Ultrasound studies. If you have not had an abnormal TCD study by your teenage years it is less likely that you will have a stroke. Ask your doctors if want to know more about strokes and your risk of having one.

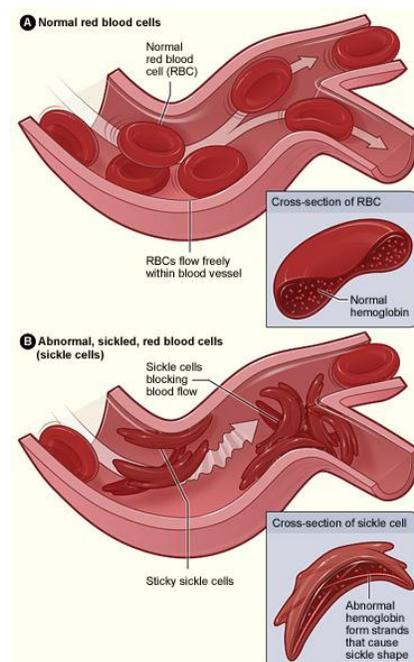
### *What causes pain?*

Patients with sickle cell disease have red blood cells that form 'sickle' or half-moon shapes (see next page). These sickle shaped cells can stick in the blood vessels, block blood flow, and cause pain. When the individual with Sickle Cell Disease is well, his or her red cells are shaped like those of people without the disease. It is when the individual with Sickle Cell Disease is stressed or sick that the cells become sickle shaped.

This vaso-occlusive pain can be triggered (caused) by factors such as cold weather, having an infection, being dehydrated, or at times of stress. You can help prevent some pain episodes by avoiding these kinds of triggers.

Some women with sickle cell disease notice they have more sickle pain around the time of their period. If this happens to you, please discuss it with your doctor. Sometimes hormone therapy may help here.

The two pictures below show the difference between normal red blood cells in the blood vessel and sickled red blood cells, and how the sickled red cells can block blood flow to organs which often results in painful episodes.



From NHLBI: <http://www.nhlbi.nih.gov/health/health-topics/topics/sca/>

### *How can I prevent pain?*

- Always dress appropriately for the weather; make sure you have warm clothes with you in case the temperature drops.
- Limit your time outside when it is very cold.
- If you go swimming, always swim in a heated pool.
- Keep yourself well hydrated - in sickle cell disease, your kidneys do not hold on to water as well, so you will need to drink extra fluids, especially if it is warm outside or when you are exercising.

### *How do I treat my pain?*



You should have an individual pain treatment plan that you have discussed with your doctor. This should include:

- drinking plenty of fluids.
- using warm packs, or taking a warm shower or bath.
- using your anti-inflammatory medicine (ibuprofen) and a short-acting opioid medicine (such as Oxycodone) when pain starts. It is OK to take these medications together, and the sooner you start treating your pain the better.

If your pain is not controlled, or if you have any questions, call your Hematologist.

Always make sure to have pain medication with you (at home, at school, or in your bag if you might be spending time away from home). If you are travelling, call your doctor to let them know, keep your prescriptions in the original bottle, and stay hydrated.

**Refills for opioids (Oxycodone, Percocet, Tylenol with Codeine, Dilaudid or Long Acting Morphine) must be given to you in person. These drugs cannot be called, sent in electronically, or faxed to the pharmacy. Therefore always make sure to contact your doctor for a refill before you run out of your medicine.**

### *Pain Medication Education*

#### **Ibuprofen:**

- pain relief and anti-inflammatory.

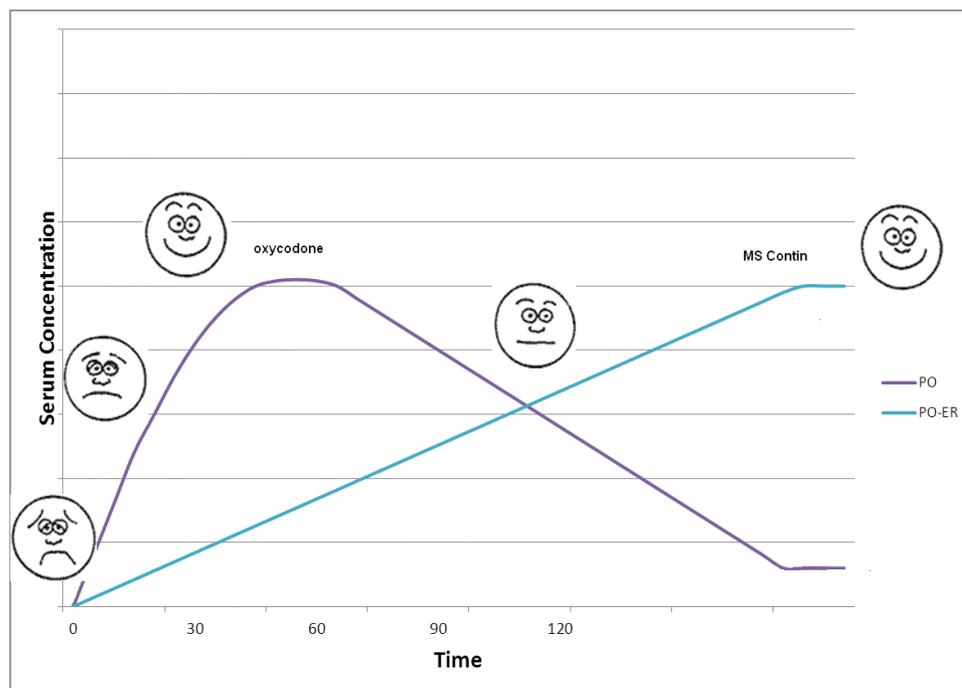
#### **Oxycodone:**

- fast acting/short lasting opioid pain medication (starts working in 20 minutes, lasts 4-6 hours).

#### **MS Contin (extended release morphine):**

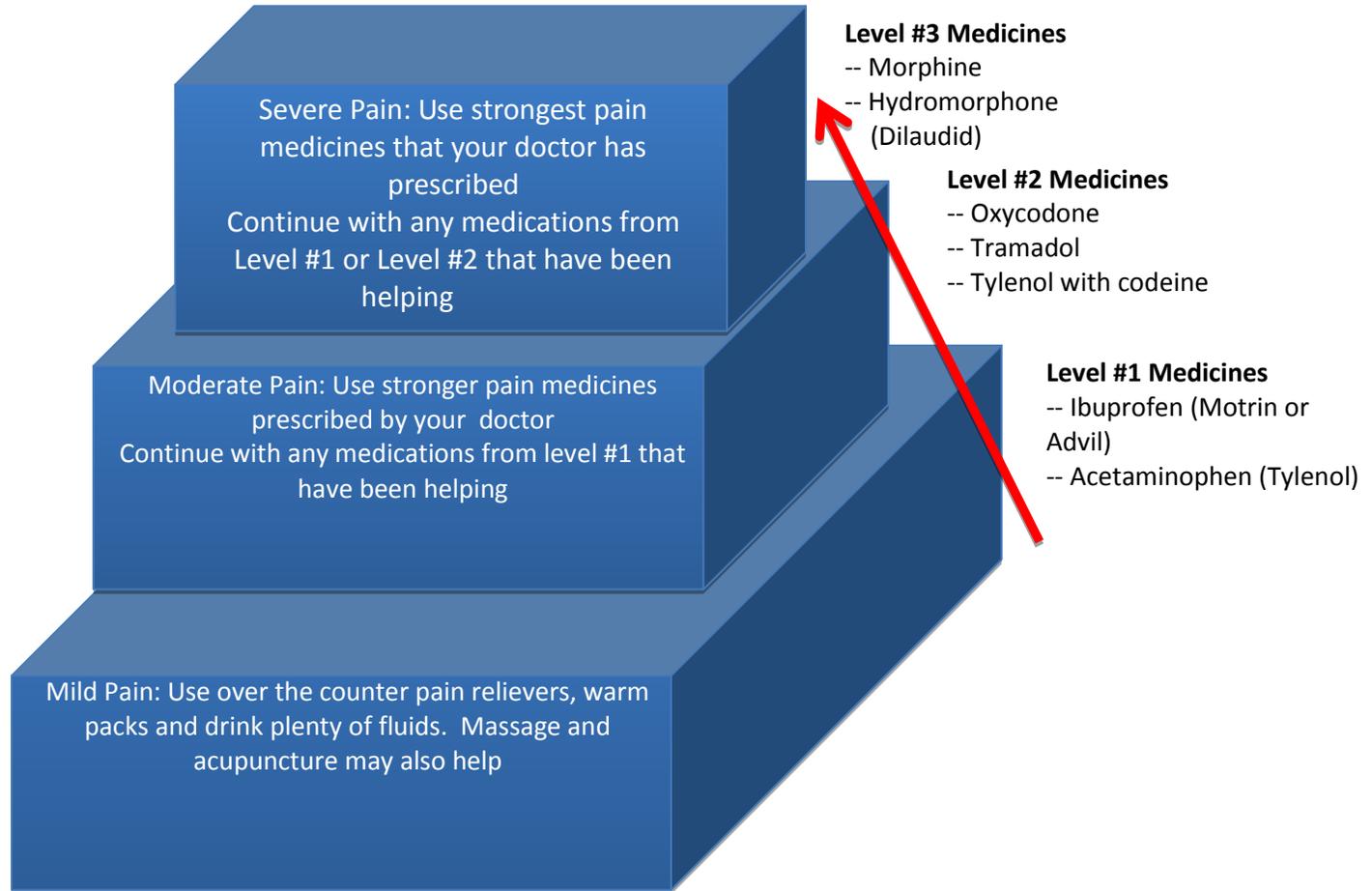
- slow acting/long lasting opioid pain medication (starts working in 1 ½ hours, lasts 8-12 hours).

It is OK to take these medications together.



\*Serum Concentration = blood level

*Pain pyramid*



**SICKLE CELL PAIN  
(WITHOUT FEVER)**

- Remember: 1.) Never run out of medicine.  
2.) Call your doctor for help before having to go to the ED

### *What if my pain is interfering with my life?*

There is a medication called **Hydroxyurea** that has been shown to reduce the number of painful episodes for many people with sickle cell disease. Studies also show that Hydroxyurea may help you live longer and healthier life with the disease. If you have frequent pain and it is interfering with work, school, etc. talk to your doctor about Hydroxyurea.

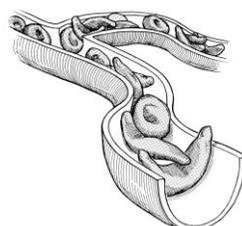
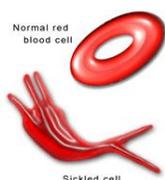
## **HYDROXYUREA FOR INDIVIDUALS WITH SICKLE CELL DISEASE**

### What is Hydroxyurea?

Hydroxyurea is a medicine that has been shown to help adults and children with sickle cell disease. Studies have shown that Hydroxyurea can decrease the number of pain events, acute chest syndrome (pneumonia), blood transfusions and admissions to the hospital. Patients take it by mouth once a day, either as a capsule or liquid. It must be prescribed by a doctor and is available at most drug stores. 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 sickle cell for close to 20 years.

### How does it work?

Red blood cells contain hemoglobin, which carries oxygen. In sickle cell patients, abnormal 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.



### Who should take Hydroxyurea?

People with sickle cell disease may be eligible for Hydroxyurea treatment. Only those who can make regular Hematology Clinic visits for required blood counts and physical exams would be given Hydroxyurea. People with severe complications of sickle cell disease (frequent pain, pneumonia, very low hemoglobin, etc.) are especially suited for Hydroxyurea. Even those with milder disease may benefit since the treatment may prevent sickle complications.

### What are the proven benefits of Hydroxyurea?

- Fewer pain events
- Fewer episodes of pneumonia and acute chest syndrome
- Fewer hospital admissions
- Fewer blood transfusions
- Improved appetite
- Increase in height and weight
- More energy
- Fewer missed school days

### Are there side effects?

Hydroxyurea is extremely well tolerated. It can cause a decrease in blood counts and must be carefully monitored for this side effect. A monthly blood count is usually required.

### Is it safe?

Yes. There have been no severe side effects seen in small groups of children, ranging in age from 6 months to 15 years. Some pediatric patients have been treated with Hydroxyurea for more than ten years, and they have no reported irreversible complications.

### How do you take it?

Hydroxyurea comes as a capsule or a liquid and is taken by mouth just once a day. Treatment will begin on a low dose daily based on weight and increases every few weeks depending on its effect.

### What is the cost of Hydroxyurea?

Hydroxyurea is fully covered by most private drug insurance policies and by MassHealth.

### How do I get more information?

Contact your Hematologist for more information about whether Hydroxyurea is right for you.

### ***Other methods of Treating Pain***

Pain can be a life-long challenge for those living with sickle cell disease. In addition to medical intervention, there are a number of non-medical approaches that may help you better cope with pain. Some of these you may be able to provide yourself and some you will need to discuss with your doctor.

For more information, please contact the Program for Integrative Medicine and Health Disparities at Boston Medical Center, on 617-414-6795.

**Acupuncture/Acupressure** 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 you are having pain. This needle blocks the meridian, to stop or decrease the pain.

Acupressure uses the same pressure points and meridians as Acupuncture, but a gentle to firm finger pressure is applied instead of needle.

**Aromatherapy** is a way of using good smells to help you 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 mood and helps you relax. It may also help your brain make special chemicals like endorphins, which can help decrease pain.

**Breathing exercises** are another physical way to help your body relax. Teaching the body to relax helps lessen pain. You should teach yourself to breathe in and out very slowly. A fun way to practice breathing slowly is to blow soap bubbles or a party blower.

**Distraction** teaches you to focus your attention on something other than pain. Watching TV, playing board games, or video games may help you relax. This can help keep you from thinking about the pain. Take these with you when you go for treatments or procedures.

**Guided imagery** teaches you to put pictures in your mind to help make the pain less intense. With guided imagery, you learn how to change the way your body senses and responds to pain. You can imagine floating in the clouds, remember a favorite place, or imagine doing a favorite activity. Think about the things you see while you are doing this.

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

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

**Relaxation and biofeedback** teach your 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 breathing also becomes faster and shallower. These responses can make the pain worse. Relaxation helps make the pain less by changing these responses.

**Massage** is often used to help someone become more relaxed. You can ask friends or family members to gently massage your back, shoulders, and neck. Massage may work even better if you use guided imagery, breathing exercises, or music.

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

**Self-hypnosis** is a way for you to change your level of awareness. This means that by focusing your attention, you can move away from your pain by making yourself 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 your normal activities. Self-hypnosis gives you better control of your body. You may feel less hopeless and helpless because YOU are doing something to decrease the pain.

### ***Can I still do the things I enjoy?***

Absolutely, just use common sense. For instance, take frequent water breaks while you are playing sports. Do not become dehydrated being outside in the heat / humidity. Make sure you have warm clothes if you like to spend time outside in cooler months.

### ***What else should I think about?***

Alcohol and caffeine can both lead to dehydration. It is advisable to drink in moderation. If you are consuming alcohol or caffeine, increase the amount of water you are drinking.

### *What is a Fever and what should I do if I develop a fever?*

Fever is a temperature of greater than normal (98.6 °F). In order to ensure adequate care, any Sickle Cell Disease individual with a temperature reaching 101.5 °F (38.5 °C) must call and seek medical advice.

Fever is a normal response of the body to infection. Remember: although taking Tylenol or Motrin may take your temperature down, these medicines do not treat the infection. You are not cured of the infection just because your temperature has come down. Always call your doctor if you have a fever of 101.5 °F or greater. Keep a thermometer at home so you can take your temperature. If you have a lower grade fever but feel sick at the same time, this is also a reason to call your doctor.

People with sickle cell disease are at higher risk of infections because their spleen does not work properly to remove bacteria from the blood. To prevent an infection, make sure you are up to date on all your vaccines, especially pneumococcal and meningococcal vaccines. The pneumococcal vaccines may protect you from developing pneumonia, an infection of the lungs and the meningococcal vaccines may protect you from getting meningitis, an infection around the brain.

Make sure you have a thermometer with you that you know how to use so that you can use it to check your temperature if you think you have a fever. Ask your doctor to write a prescription for one if you don't have one.

If you have a fever, come to the emergency room right away. At the ER, they will take a sample of blood and test it for any bacteria. Since bacteria can take 1-2 days to grow they will give you a dose of antibiotics while waiting for results. In many cases you will be able to go home from the ER.



### *How do I meet my daily fluid needs?*

Individuals with sickle cell disease have a higher risk of developing dehydration than other people because their kidneys cannot hold water. It is important therefore to drink plenty of fluids to meet your daily fluid needs and prevent dehydration. Dehydration may cause your red blood cells to sickle more than normal and this can lead to painful episodes.

We advise you:

- carry a water bottle with you at all times.
- start your day by drinking a glass of water and then have something to drink approximately every 2 hours.
- drink extra fluid when you have pain or with exercise.
- choose fluids such as water (spring/tap/flavored), milk, and 100% fruit juice over others (soda, coffee, tea or fruit drinks).
- drink about 12 cups of water per day.

### *Eating Healthy Diet – Do I have to go on a special diet?*

There is no special diet for anyone with sickle cell disease. However, it is important to eat a healthy, balanced diet including fruits, vegetables, whole grains, fat-free or low fat milk or milk products, lean meat and fish.

Studies have revealed that individuals with sickle cell disease need about 20 percent more calories than others to fuel their production of red blood cells. Not getting enough calories may lead to delays in growth and maturation.

It is advisable to eat less saturated fats, trans fat, cholesterol, sodium and added sugar.

This table shows the different food groups that make a balanced diet

Food Groups	Examples of foods and what they give our bodies.
<b>Grains</b> 	<p>Whole grain breads, rice, pastas, cereals and crackers – aim to make at least half of the grains that you eat “whole” grains</p> <ul style="list-style-type: none"> <li>• Fiber – helps to reduce blood cholesterol and keeps your bowels moving</li> <li>• Folate (in fortified grains) – helps our body to make new red blood cells</li> <li>• B-vitamins – helps our body to use the energy from foods that we eat</li> <li>• Magnesium – important for healthy bones</li> <li>• Selenium – important for healthy immune system</li> </ul>
<b>Vegetables</b> 	<p>Spinach, carrots, broccoli, tomato, green beans, salad – aim for orange and dark green leafy vegetables</p> <ul style="list-style-type: none"> <li>• Fiber – Tip: keep the skin on veggies and fruits for extra fiber</li> <li>• Folate – Tip: choose leafy greens</li> <li>• Vitamins A – keep eyes and skin healthy, helps protect against infections</li> <li>• Vitamin C – helps heal wounds, keeps teeth and gums healthy</li> <li>• Potassium – may help maintain healthy blood pressure</li> </ul>
<b>Fruits</b> 	<p>Orange, bananas, apples, grapes, melon, berries, kiwi, peach, plums</p> <ul style="list-style-type: none"> <li>• Provide similar vitamins, minerals and fiber as vegetables</li> <li>• Note: individual fruits and vegetables provide different amounts of vitamins, minerals and fiber so it is important to eat a variety of different fruits and vegetables daily to maximize your benefits.</li> </ul>
<b>Milk</b> 	<p>Low fat milk, yogurt and cheese</p> <ul style="list-style-type: none"> <li>• Calcium and Vitamin D – for strong, healthy bones</li> <li>• Potassium – especially in milk and yogurt</li> </ul>
<b>Meats and Beans</b> 	<p>Lean chicken, pork, fish and beef, nuts, seeds, peas and beans</p> <ul style="list-style-type: none"> <li>• Protein and zinc – for energy and growth</li> <li>• Iron – helps to carry oxygen in our blood</li> <li>• Omega 3 fatty acids – in certain fish, nuts and seeds – help to reduce inflammation and decrease risk of chronic diseases, like heart disease</li> </ul>
<b>Oils</b> 	<p>Liquid vegetables oils like olive, canola, corn oil; nuts</p> <ul style="list-style-type: none"> <li>• Vitamin E – works as an antioxidant to help to prevent disease</li> <li>• Omega 3 fatty acids – in flax, canola, soybean and walnut oils, and walnut flaxseeds and pumpkin seeds</li> </ul>

### *Does Sickle Cell Disease affect pregnancy in any way?*

Most commonly, sickle cell does not affect fertility and many young women with sickle cell disease have had successful pregnancy outcomes.

Women who may become pregnant, or men who may father a child, should talk to their hematologist if they are on Hydroxyurea, since they may need to stop the medication while pregnant.

Sickle cell disease can also worsen during pregnancy. There is an increased chance of having pre-term birth, low birth weight babies, and pregnancy induced hypertension. Therefore women with sickle cell disease should get special prenatal care and frequent checkups to ensure a healthy pregnancy.



## Contraception (Safe Sex)

### Things to Remember:

- Becoming sexually active and with whom is a big decision.
- Having sex should be YOUR choice and no one should pressure you.
- Wait until you are ready and protect yourself against unwanted pregnancies & sexually transmitted diseases (STDs)
- Decide on how to prevent pregnancy and STD *before* becoming sexually active.
- Contraception methods vary in effectiveness and cost.
- Using two methods (such as birth control pills and condoms) greatly decreases your chances of becoming pregnant and getting an STD.



As a teenager, you may be starting to think about what it means to be involved in a sexual relationship. There are many things that you need to think about before you decide to have sex, including whether this is the right person, the right time in your life, and how you will feel if the relationship breaks up. If you do decide to have sex, you definitely need to think about how to prevent getting pregnant and how to protect yourself from getting a sexually transmitted disease (STD). Have a discussion to decide what type of contraception is best for you and your partner. You should talk to a trusted adult or your health care provider if you are thinking about having a sexual relationship. It is a good idea to discuss all of your choices and all of the concerns and worries you may have so you can make good decisions. This can be a very confusing time for you and it is always good to have someone to talk to.

## What types of contraception are there?

From [www.youngwomenshealth.org/conta.html](http://www.youngwomenshealth.org/conta.html)

<p><b>Birth Control Pills</b></p> <p><b>Minimum effectiveness: 95%</b></p>	
<p><b>Pros</b></p> <p>Very effective against pregnancy if used correctly            Makes menstrual periods more regular and lighter            Decreases menstrual cramps and acne            Makes you less likely to get ovarian and uterine cancer, pelvic inflammatory disease, ovarian cysts, and anemia            Doesn't interrupt lovemaking            Should use condoms to lower the risk of STDs</p>	<p><b>Cons</b></p> <p>Doesn't protect against STDs            Need to remember to take every day at the same time            Can't be used by women with certain medical problems or by women taking certain medications            Can occasionally cause side effects such as nausea, increased appetite, headaches, and, very rarely, blood clots            Need a prescription</p>
<p><b>Vaginal Hormonal Ring (Nuva-Ring)</b></p> <p><b>Minimum effectiveness: 98%</b></p>	
<p><b>Pros</b></p> <p>Very effective against pregnancy if used correctly            Makes menstrual periods more regular and lighter            Decreases menstrual cramps and acne            Makes you less likely to get ovarian and uterine cancer, pelvic inflammatory disease, ovarian cysts, and anemia            Doesn't interrupt lovemaking</p>	<p><b>Cons</b></p> <p>Doesn't protect against STDs            Should use condoms to lower the risk of STDs            Can't be used by women with certain medical problems or by women taking certain medications            Can occasionally cause side effects such as nausea, increased appetite, headaches            Higher risk of blood clots            Need a prescription</p>
<p><b>Hormone Patch (Ortho-Evra)</b></p> <p><b>Minimum effectiveness: 99%</b></p>	
<p><b>Pros</b></p> <p>Very effective against pregnancy if used correctly            Makes menstrual periods more regular and lighter            Decreases menstrual cramps and acne            Makes you less likely to get ovarian and uterine cancer, pelvic inflammatory disease, ovarian cysts, and anemia            Doesn't interrupt lovemaking</p>	<p><b>Cons</b></p> <p>Doesn't protect against STDs            Should use condoms to lower the risk of STDs            Can't be used by women with certain medical problems or by women taking certain medications            Can occasionally cause side effects such as nausea, increased appetite, headaches            Higher risk of blood clots            Need a prescription</p>

<p><b>Depo-Provera Hormonal Injection</b></p> <p><b>Minimum effectiveness: 99%</b></p>	
<p><b>Pros</b></p> <p>Each injection provides 3 months of protection against pregnancy          Very effective against pregnancy          Many women stop getting their menstrual period while getting injections. (This is not a medical problem and menstrual periods usually return 6-18 months after you stop taking injections)          Helps protect against uterine cancer</p>	<p><b>Cons</b></p> <p>Doesn't protect against STDs          Need to see your health care provider every 3 months for an injection          May have side effects such as weight gain, tiredness, and possibly a decrease in bone density          Many women have very irregular menstrual bleeding or spotting for the first 3 to 6 months and sometimes longer</p>
<p><b>Male Condom</b></p> <p><b>Minimum effectiveness: 86%</b></p>	
<p><b>Pros</b></p> <p>Lowers risk of STDs          Effective against pregnancy          Contraception that provides the most protection against sexually transmitted diseases (latex condoms are best)          Doesn't cost much (50 cents each), can buy at almost any drug store (don't need a prescription)          Allow men to have an active part in preventing pregnancy</p>	<p><b>Cons</b></p> <p>Have to use a new one every time you have sexual intercourse (can only be used once)          May disrupt/interrupt lovemaking          Can break          Women may be allergic to latex</p>
<p><b>Intra-Uterine Device (IUD)</b></p> <p><b>Minimum effectiveness: 99%</b></p>	
<p><b>Pros</b></p> <p>Very effective against pregnancy          Provide protection against pregnancy as long as in place in your uterus- protects as soon as inserted          Doesn't need daily attention- just need to check to make sure in place at least once a month at time of menstrual period          Comfortable- you and your partner cannot feel the IUD, although you partner may feel the string          The levonorgestrel IUD (Mirena) lessens menstrual flow and can be used to treat heavy periods          Can be removed at any time</p>	<p><b>Cons</b></p> <p>Doesn't protect against STDs          Needs to be inserted by a health care provider          It is best for women who have already had children and are in a steady relationship with one partner.          Can fall out or can rarely puncture the uterus          The copper IUD can have side effects such as menstrual cramping, longer and/or heavier menstrual periods, and spotting between menstrual periods          Slightly higher risk for infection in the few weeks after insertion</p>

### ***Dental Care.***

Always schedule and keep appointments with your dentist in order to prevent mouth or teeth infection that may lead to complications of sickle cell disease. A dental evaluation should be carried out every 6 months.

Cleanings and dental fillings do not require special care, but operative procedures such as extractions and root canals may require antibiotic prophylaxis in order to reduce the risk of bacterial infection. Dental procedures under anesthesia may require a transfusion ahead of time. If you need dental work done, make sure your dentist and Hematologist discuss your case before anything is done.



### ***Should I be worried about taking too much medicine or developing an addiction to medications?***

No! If you use your pain medications appropriately as prescribed by your doctor, it will not lead to addiction.

Make sure you follow the instructions as to when and how to take pain medicines given by your provider(s) and take the required doses to control your pain. If your pain persists despite all medications given to you, please call your doctor or report to the ER. Never use your medications to relief stress or anxiety. Always ask for medical advice if you need help with stress or anxiety.

### ***What about Cigarettes, Alcohol and illicit drugs?***

Cigarette smoking harms nearly every organ in the body. As someone living with sickle cell disease, the chemicals in the tobacco smoke may harm your blood cells.

Tobacco can also damage the structure and function of your blood vessels, as well as the function of your heart and lungs. This damage will increase your risk of developing atherosclerosis (a disease in which a waxy substance called plaque builds up in the arteries). The plaques harden overtime and destroy the blood vessels. These plaques, in addition to blockage caused by sickled hemoglobin, may lead to frequent pain crises and organ damage.

Also try to avoid second hand smoke. Ask friends and family members who smoke to quit, or not to do it in the house and car.

If you are smoking, we advise that you quit. If you are ready to quit, please contact your primary care provider or a community program for resources that will be useful for you.

Alcohol will cause dehydration which can predispose you to pain crises. If you do want to drink, please do so in moderation and never take so much that you lose control of your own wellbeing.

Ecstasy and Marijuana (Cannabis) have been associated with frequent incidence of priapism in patients with sickle cell disease.

### *Tattoos and body piercings*

Having Tattoos on your body may increase your risk of getting an infection if the tools are not sterilized. Make sure you are going to a reputable and licensed place with clean equipment.

### *Can I participate in sports?*

Your doctors will make every effort to support you in any physical activity you would like to undertake. There are sports that may be safer and others that may be less safe for someone with sickle cell disease. We strongly advise that you go for an assessment to decide which would be the best activities for you.

As a general rule, any sport that would require overexertion, causes overheating, chilling or dehydration may trigger pain crises. In addition to this, any form of activity in a high altitude could also be harmful to you.

If you have an enlarged spleen, it is advisable you avoid contact sports which may predispose the spleen to rupture.



## Coping with Sickle Cell Disease

As a young adult, you are beginning to see yourself as an individual in the world while you strive to think and act independently. Living with sickle cell disease may cause a huge stress on you emotionally and otherwise. It makes you different from your friends/peers, and you may get concerned with how they view your illness.

There are many ways to cope with the stress of the disease. Seek advice and ask questions from your providers during visits to the hospital/clinic, talk with families, friends and others who are facing the same challenge as you. Prayer is also a potential source of support.

### *Tips for supporting yourself when you are ill or hospitalized:*

- Remain active in social and school activities as you are able.
- Be actively involved in your treatment plan by talking to the team and providers.
- Allow yourself to do things that make you feel good about yourself.
- Let those around you know what you are capable of doing on your own and what things you might need some help with.
- Let your family and friends visit or call you in the hospital or home.
- Ask for help to plan for the future.
- Keep doing normal things like school and work.
- Find ways to express your feelings such as physical activity, talking, or writing. Know that all of your feelings are normal, such as guilt, fear, or sadness. However, if these occur all the time, please call your doctor.



## Sickle Cell and Your Education/Career

Thinking of going to college?

- Create list of schools that you are interested in going to/finding more information on.
- Track deadlines (when are applications due? e.g. for FAFSA – Free Application for Federal Student Aid), and keep on top of teachers/mentors who are completing letters of recommendations.
- Apply for other loans, grants, etc.
- Fill out college applications, complete FAFSA applications for financial aid.
- Talk with guidance counselors at your high school.
- Talk with your Social Worker at the Transition Clinic



## Special Education



Individuals with SCD often face many challenges during school due to frequent absence from school. In order to overcome these barriers and have a successful educational experience, the state and federal government have legal, written documents that are centered on your specific needs in school.

Children with disabilities or special needs and education between the ages 3-21 years benefit from laws that advocate for accommodations, special instructions and services they receive in school.

Some of these laws include the Individualized Education Program (IEP) and the 504 Plan.

### Individualized Education Program (IEP)

The Individualized Education Program describes special educational services and accommodations. This program must be reviewed every year and should include the following:

- A list of your strengths and areas to improve.
- A vision statement of your hopes and goals.
- Measurable educational goals and a plan for achieving them.
- Specially designed teaching and/or services (such as speech, occupational, and physical therapies, assistive technology) you need to reach those goals.
- Beginning at age 14, a description of special instruction that will help you transition from school to adult life activities when ready.

## 504 Plan

A 504 plan helps you to fully participate in the general classroom setting and educational programs. It includes:

- Extra sets of textbooks.
- Access to tutors when hospitalized.
- Classroom liaison to communicate missed or catch-up work.
- Hall passes to visit the restroom.
- Ability to wear coats when necessary.
- Elevator access.
- Modified or limited participation in gym.
- Plans to make a school wheelchair accessible.
- Your assistive technology needs during the school day.
- Permission to type assignments instead of writing them by hand.
- Permission to hand in assignments late due to illness or a hospital stay.

## Individualized Health Care Plan (IHCP)

An IHCP helps to make sure you receive the health services you need during the school day. Although there is no law that says you have a right to an IHCP, you can ask for an IHCP if you need one. Contact your school nurse and your primary care provider.

The IHCP should include:

- The health services you receive at school.
- When, where, and how the health services will be provided.
- Who will provide the health services.
- Information about your transportation needs, including special needs for field trips.
- An Emergency Response plan that lists possible emergencies and what to do.
- A copy of the Emergency Information Form for individuals with special health needs.

## College Support Services

### How does chronic pain interfere with Learning?

A student experiencing pain may find it difficult to perform physical skills, such as sitting, holding and using a pen. Concentrating during class or a test may be difficult. Medication may interfere with focusing or even active classroom participation. By working with your school's available support service coordinators, you can create a plan that works for your educational success! You must go to the Office of Disability at your college when you start there to let them know of your Sickle Cell Disease and have arrangements with your teachers to allow you to be excused from classes, written assignments, or exams when appropriate.

**Assumption College**

Disability Services  
P: 508-767-7500

**Emerson College**

Disability Services  
P: 617-824-8592

**Ben Franklin Institute of Technology**

Disability Services  
P: 617-588-1378

**Fisher College**

Student Accessibility Services  
P: 617-670-4429

**Boston College**

Disability Services Office  
P: 617-552-3470

**Fitchburg State University**

Disability Services  
P: 978-655-3427

**Boston University**

Student Support Services  
P: 617-353-3658

**Framingham State University**

Academic Support Services  
P: 508-626-4906

**Bunker Hill Community College**

Support Services  
P: 617-288-2234

**Newbury College**

Disability Support Services  
P: 617-730-7000

**Bridgewater State University**

Disability Resources  
P: 508-531-2194

**Massasoit Community College**

Academic Resource Center  
P: 508-588-9100 ext. 1082

**Curry College**

Student Disability Office  
P: 617-333-2385

**Mass Bay Community College**

Disability Resources  
Framingham 508-270-4267 / 4286  
Wellesley 781-239-2234 / 2626

**Massachusetts College of Pharmacy  
+ Health Sciences**

Disability Support Services  
P: 617-879-5925

**Lesley College**

Center for Academic Achievement  
P: 617-349-8194  
F: 617-349-8558

**Northern Essex Community College**

Learning Accommodations Center  
P: 978-556-3654  
P: 978-556-3654

**Mount Ida College**

Student Affairs  
P: 617-928-4563

**Quincy College**

Office of Disability Services  
P: 617-984-1656

**Lasell College**

Edward Student Center  
P: 617-234-2451

**Middlesex Community College**

Disability Support Services  
P: 978-656-3258 9 (Lowell)  
P: 781-280-3630 (Bedford)

**Regis College**

Disability Support Services  
P: 781-768-7384

**Pine Manor College**

Brown Learning Resource Center  
P: 617-731-7181  
F: 617-731-7638

**Salem State University**

Disabilities Services  
P: 978-542-6217  
F: 978-542-2064

**Northeastern University**

Disability Resource Center  
P: 617-373-2675  
F: 617-373-7800 (confidential)

**Roxbury Community College**

Students with Disabilities Center  
P: 617-427-0060 ex. 5006

**Simmons College**

Disability Services

P: 617-521-2472

F: 617-521-3079

**UMASS Dartmouth**

Center for Access and Success

P: 508-999-8711

**UMASS Lowell**

Student Disability Services

P: 978-934-4574

F: 978093404574

**Wheelock College**

Ross Center for Disability Services

P: 617-287-7430

**Westfield State**

Banacos Academic Center

P: 413-572-5676

**Worcester State**

Disability Service Office

P: 508-929-8733

F: 508-929-8214



## ***Finding a Job***

First time job searching? What would you like to do for work?

- Fill out applications either on site or online
- Put together a resume.
- Do you own appropriate dress attire?
- Prepare for the interview.

Second, third, fourth ...twelfth job?

- Keep your resume up-to-date.
- Write a cover letter.
- Keep a positive attitude.
- Make sure your wardrobe is appropriate.
- Practice job interview skills with a friend or even in the mirror.

Information on jobs, careers, and training can be found on:

<http://www.usa.gov/Citizen/Topics/Education-Training/Finding-Jobs.shtml>

<http://bostonjobsource.com/>

<http://www.monster.com/>

<http://www.simplyhired.com/>

<http://www.indeed.com/>

<http://www.careerbuilder.com/>

<http://www.snagajob.com/>

### *Are there Job accommodations for people with sickle cell disease?*

Yes! It is important for you to inform your future employer of your medical condition before taking up any job. The following guidelines on job accommodation have been put together by the Job Accommodation Network (JAN), and the U.S. Department of Labor's Office of Disability Employment Policy.

Because individuals with sickle cell disease may have a recurrent need for medical intervention or transfusions, your employer should:

- allow time off for medical appointments.
- allow you work from home full time, part-time, or as needed.
- permit a family member or friend to call-off for you when in "crisis".
- provide additional unpaid leave if you exhaust accrued time off.
- allow telephone calls during work hours to doctors and others for support.
- provide information on counseling and employee assistance programs.

Secondly, to avoid fatigue or weakness while at work, your employer can allow you:

- schedule periodic rest breaks away from the workstation, and reduce physical exertion and workplace stress.
- design a workstation that best suits your body (Ergonomics).

Thirdly, to prevent respiratory difficulties / reduced immunity, your employer can:

- provide adjustable ventilation and air cleaner.
- keep work environment free from dirt, dust, smoke, odors, and fumes.
- provide anti-bacterial wipes to clean workstations.
- implement an illness policy, requiring employees to stay home if contagious to avoid exposing co-workers to colds, flu or other air-borne infections.

Lastly, your employer can modify work-site temperature or provide you an office with separate temperature control. You can also be allowed to use fan/air-conditioner or heater at the workstation. For further information on job accommodation, please visit [www.askjan.org](http://www.askjan.org).

## Health Insurance

As a young person with a chronic illness, it is very important that you have health care coverage for your doctor's visits, medications, and hospitalizations.

As a Massachusetts resident, it is the law that you must have insurance (this is called an **individual mandate**). There are several kinds of health insurance out there and the process for getting and staying on insurance can get complicated. Here is some basic information to guide you.

### Public Programs

Public programs are health insurance programs paid for in part by the state and federal governments.

**MassHealth** is insurance for Massachusetts residents. The level of MassHealth coverage that one is eligible for is based on income and disability. MassHealth services may be limited for people who are NOT disabled, and depending on immigration status. For more information, go to:

[www.mass.gov/eohhs/gov/departments/masshealth](http://www.mass.gov/eohhs/gov/departments/masshealth)

**Commonwealth Care** is an individual insurance plan offered by the state. Individuals who cannot get MassHealth or coverage through their school, work, or parents can purchase a plan through the Massachusetts Connector Board. The state will sometimes subsidize (help pay for) the cost of this insurance depending on your income. For more information, go to:

[www.mahealthconnector.org](http://www.mahealthconnector.org)

**Medicare** is a federal health insurance program for people age 65 and older OR who have a permanent disability and have been on SSI/SSDI for at least two years. For more information, go to: [www.medicare.gov](http://www.medicare.gov)

## Private Programs

These are health insurance policies paid for by individuals and/or employers:

**Through Mom and Dad:** If your parents have a health insurance plan through their employer, their child can stay on the plan “for up to two years after you lose **dependent** status, up to the age of 26 years.”

**Through Work:** If your employer offers a plan, you usually can sign up when you are hired (you may have to wait for 30 days for coverage to kick in). If you do not elect a plan at the time of hire, you might have to wait until an open **enrollment period** to get coverage.

**Through School:** Many colleges require students to have a health insurance plan. This can be your family plan, MassHealth, Commonwealth Care, or a plan offered through the school.

## Dental Coverage

As with regular health insurance, there are different dental insurance programs that you may be eligible for.

**MassHealth Dental Coverage:** If you are on MassHealth, they will cover emergency and preventative (check-ups) treatment for adults over the age of 21. Adults over 21 do NOT have coverage for “restorative” services, like fillings, root canals, crowns, dentures, etc.). Emergency, preventative, and restorative treatment is available if you are under 21 and on MassHealth.

**Other Dental Health Care Plans:** You may be offered dental insurance through your school, work, or parent’s insurance plan. Dental insurance is not mandatory in Massachusetts.

There are several clinics offering reduced-cost dental care. Find more info at: <http://www.massdental.org/content.aspx?id=792>.

## Your responsibilities

It is important to have continuous health care coverage so that you can access your doctors, use emergency services, and keep your prescriptions filled. As mentioned above, in Massachusetts if you do not have health insurance you can be fined when you do your tax returns. Here are some things to remember when it comes to keeping your insurance:

- Keep your insurance company updated if your address or phone number changes.
- Answer any mail sent to you by your health insurance company. MassHealth sends a yearly **“eligibility review”** form that must be completed and returned, otherwise your MassHealth coverage will end.
- If you have Commonwealth Care or a private insurance plan, you must pay your **premiums** and **deductibles** in order not to lose coverage.
- Your insurance company will send you a statement (by mail or email) every year stating that you have **“minimum creditable coverage”**. A copy of this statement needs to be sent in with your tax returns so you can show the state that you have complied with the mandatory health insurance law.

**As always, if you have any questions about health insurance, call your social worker in the sickle cell clinic for help!**

### Important Health Insurance Terms:

**Copay:** A predetermined (flat) fee that an individual pays for health care services, like medications, in addition to what the insurance covers.

**Deductible:** The amount an individual must pay for health care expenses before insurance (or a self-insured company) covers the costs.

**Dependent:** A person or persons relying on the health insurance policy holder for support. This may include the spouse and/or unmarried children (whether natural, adopted, or step) of the health insurance policy holder.

**Eligibility Review:** MassHealth's yearly process of reviewing its members' eligibility for MassHealth insurance.

**Enrollment Period:** The time during which you are allowed to enroll in a group health insurance plan. As a rule, there are specific times when others may join the group or change their existing coverage. Those times vary depending on the insurance.

**Individual Mandate:** Massachusetts residents age 18 and older must have health insurance.

**Minimum Creditable Coverage:** The essential health insurance benefits you need to avoid tax penalties in Massachusetts.

**Premiums:** The money paid by an insurance person or business for a health insurance policy.

**Subsidy:** Help with the cost of health insurance premiums, most often offered by the state or federal government.

## Income Supports for Persons with Disabilities

### Supplemental Security Income Benefits (SSI)

SSI provides a monthly benefit to help cover costs of living to disabled, blind or those 65 and over receiving a limited income. SSI is not Social Security, but is administered via the Social Security Administration. To find out eligibility requirements and apply, visit [www.ssa.gov/ssi](http://www.ssa.gov/ssi).



## Social Security Disability Insurance (SSDI)

SSDI provides a monthly stipend to help cover costs if you are disabled and unable to work. SSDI is a part of Social Security and requires that applicants have contributed to social security while employed. SSDI is given for as long as the individual is disabled and unable to work due to a medical condition. Generally, Social Security determines that someone is disabled if they cannot work for one year or longer.

Visit [www.ssa.org/disability/](http://www.ssa.org/disability/).

Other sites include:

<http://www.ssa.gov/boston/>

<http://www.ssa.gov/boston/MA.htm>

<http://socialsecurityhop.com/offices/social-security-office-boston-ma-02119-massachusetts>

[http://www.ultimatedisabilityguide.com/ma\\_ssd\\_ssi.html](http://www.ultimatedisabilityguide.com/ma_ssd_ssi.html)

## People who can help you make decisions about your health, financial, and personal affairs

- **Guardian**– If due to a medical condition, such as a stroke, an adult is not *able* to make *effective* decisions about their own *care, daily needs, and safety*, a guardian can be appointed by the Massachusetts’ Court to help make those decisions on behalf of the adult in question, called the Incapacitated Person. The Guardian can also be a person previously selected by the Incapacitated Person (like a parent or family member designated as a Health Care Proxy), or an organization designated by the Court. As a Guardian, that person or organization makes key decisions about the Incapacitated Person’s well-being, care, and daily needs while respecting their autonomy and personal rights. A Guardian can be temporary or have limited authority depending on a person’s needs. For more information visit [www.massguardianshipassociation.org](http://www.massguardianshipassociation.org).
- **Representative Payee**- This is an individual, such as a family or friend, designated to help SSI and SSDI recipients manage their funds if they are incapable due to a medical condition or in emergencies. If a person can not be appointed, the Social Security Agency can designate an individual of their choosing. This is different from a Guardian, Conservator and Health Care Proxy. For more information, visit [www.socialsecurity.gov/payee/](http://www.socialsecurity.gov/payee/)
- **Health Care Proxy** – This legal document, signed by the patient, designates an individual (the “agent”) to help make health care decisions for the patient *if they are unable to communicate with the staff*. The document is completed *prior* to any emergency, and can only be “enacted” (or activated) by a doctor *if* the patient is unable to make healthcare decisions. Visit [www.massmed.org](http://www.massmed.org) and click on “Patient” in the toolbar at the top of the screen to learn more and see a copy of the Massachusetts form.
- **Conservator of an Adult** – This is NOT a Guardian. A Conservator is a person appointed (by patient or Massachusetts Probate and Family Court) to provide *protection* in managing *money, property or business* for a patient (called the Protected Person) who is unable to do so due to a disability. Visit [www.massguardianshipassociation.org](http://www.massguardianshipassociation.org) for information.

## Independent Living

### Housing

The first thing you should know before getting your own apartment is to understand your rights as a tenant, as well as knowing the rights of your landlord.

According to the law, a tenant has the right to:

- Use the property they are renting according to the terms of lease.
- Privacy. Your landlord may only enter your apartment to make repairs, deliver a message, inspect or show the apartment to anyone who intends to move in.
- File complaints against your landlord if the conditions of the apartment become a threat to your health or safety.
- Know the contact information of your landlord.

On the other hand, you should realize that your landlord also has the right to:

- Have the property maintained by you according to the terms of the lease.
- Rent the apartment to anyone of his/her choice, but must not discriminate according to race, color, religion, sex, nationality, or disability.

### *What is a Lease?*

A lease is an agreement or contract between a landlord and a tenant. It is a legal document that protects the rights of both you and the landlord. Before signing a lease, always read carefully to make sure you are willing to abide by everything written in it. Take note of the length of time (months/years) the lease covers, how much the security deposit is, who pays the utility bills, the day rent is due, and other Dos and Don'ts while living in the apartment.

Some landlords require a security deposit before moving in. This in most cases amounts to a month's rent and often used to cover any damage you may cause while living in the apartment. You must also keep the apartment clean and in good order.

### ***What are my responsibilities as a tenant?***

Once you sign a lease, it forms a legal agreement between you and the landlord. By signing the lease both you and the landlord are responsible for keeping the terms and conditions on the lease.

You should pay your rent on time. You should also obey laws related to health and safety such as garbage disposal and fire safety. However, public hallways, sidewalks, and laundry areas are to be maintained by the landlord. You should also use the apartment only for legal purposes.

### ***Where is the best place to live?***

Identify the type of house and neighborhood that best suits your needs. Also consider looking for apartment that is close to family, friends, school, work, church, laundry facilities and other community resources.

There are many resources to find housing in Boston area, some of which include:

<http://www.cityofboston.gov/rentalhousing/>

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

[http://portal.hud.gov/hudportal/HUD?src=/program\\_offices/public\\_indian\\_housing/programs/hcv](http://portal.hud.gov/hudportal/HUD?src=/program_offices/public_indian_housing/programs/hcv)

<http://www.mass.gov/hed/economic/eohed/dhcd/>

[http://www.eap.partners.org/WorkLife/Housing/Emergency\\_and\\_Low\\_to\\_Moderate\\_Income\\_Assistance.asp](http://www.eap.partners.org/WorkLife/Housing/Emergency_and_Low_to_Moderate_Income_Assistance.asp)

[http://www.eap.partners.org/WorkLife/Housing/Emergency\\_and\\_Low\\_to\\_Moderate\\_Income\\_Assistance.asp](http://www.eap.partners.org/WorkLife/Housing/Emergency_and_Low_to_Moderate_Income_Assistance.asp)

<http://www.prohope.org/programs.htm>

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

[http://portal.hud.gov/hudportal/HUD?src=/topics/rental\\_assistance](http://portal.hud.gov/hudportal/HUD?src=/topics/rental_assistance)

<https://www.masshousing.com/portal/server.pt?>

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

***Boston Center for Independent Living (BCIL)***

Promoting Independence for People with Disabilities

BCIL is a 501(c)(3) non-profit organization that has provided services to people with disabilities since the organization's founding in 1974, when it became the second independent living center in the country. BCIL aims empowering people with disabilities with the practical skills and self-confidence to take control over their lives and become active members of the communities in which they live.

BCIL can help you:

- Learn options you have for personal care and in-home support as you plan to leave a nursing home or hospital
- Find affordable and accessible housing
- Learn how you can acquire assistive technology and communication equipment
- Plan your education, particularly as you near high school graduation
- Obtain and direct PCA services
- Find out about accessible transportation services
- Obtain and maximize benefits
- Set career goals in a current or future job, or finding employment
- Address communication needs due to a language or communication barrier
- Understand your civil rights under the ADA or Fair Housing law
- Appeal a decision from housing, educational, medical provider or employer
- Increase your access to necessary healthcare

60 Temple Place, Boston, MA 02111 Phone: 617.338.6665 Fax: 617.338.6661

## Managing Money

Once you have a steady income, the next thing to do is to learn how to spend it wisely. Managing money is one of the most important skills to have as a young adult living independently.

Always keep track of your money, plan for purchases ahead of time, buy the things you need and make sure you develop a saving habit.

In addition, set up a budget for yourself (daily, weekly, or monthly) and know the difference between your “wants” and “needs”

Some common “**needs**” include: rent, water, heat, electricity, gas, food, cell phone bills, medications, personal care items, and emergency money.

Some common “**wants**” include: extra clothes, shopping money, money to go out with friends.

### *How do I use Bank Checks/ATM cards?*

First you need to open an account with a bank of your choice. A check is often used for a checking account. It cannot be used for a savings account.

A saving account is used to store money and earn interest, while a checking account provides you with an alternative to cash through the use of checks. You can write a check to pay your bills or settle other expenses.



## Automated Teller Machines (ATMs)

These are machines that can collect and also give out money in either checking or saving accounts. They are convenient, and easy-to-use. Once you have an ATM card issued to you by your bank, it can be used to withdraw and deposit money as soon as it is activated.

Note that with an ATM card, there is a greater temptation to spend money on things you never planned for. Always use your budget to guide your spending.

The use of an ATM card requires you to have a password before making a transaction at the machine. Choose a secret password, do not write it on your card or anywhere people could see it, and do not give your password to others. Always remember to remove your card from the machine at the end of every transaction. Call your bank immediately if you lose your card.



## Home Management

- Clean your living areas.
- Keep dishes clean.
- Attend to trash/recycling.
- Be aware of when your monthly bills are due.
- Going to a Laundromat? You'll need quarters and your own detergent!

## Time Management

- Plan a daily/weekly schedule.
- Make a "To-Do-List".
- Going to classes and/or to work? Try to go to bed at the same time each night and try to wake up at the same time every morning.
- Complete any necessary homework and/or work tasks.
- Set aside time for self-care (examples: going to the gym, relaxing, reading, listening to music, creating artwork, writing, practicing deep breathing exercises, etc.)
- Spend time with friends.

## Health & Safety

- Take medications as needed and at the correct times/doses/intervals.
- Schedule and go for your doctor's appointments and go for your appointments.
- Schedule follow-up appointments
- Call to cancel appointments (if necessary) at least 24 hours in advance
- Have a list of contact numbers in case of an emergency
- Take care of yourself when you have a cold/flu
- Know when to go to the hospital in a crisis.

## Transportation

### Have a car?

- Always have your license on you when you're driving.
- Make sure there is gas in the tank.
- Make sure the car registration is up to date.
- Change your oil every 3000-5000 miles.
- Make sure you have car insurance.
- AAA or other roadside assistant services can be helpful.

### Take public transportation?

- Be knowledgeable of MBTA train and bus schedules.
- Plan the route from starting to end destination.
- Be aware of bus and train stops.
- Get a Charlie card (weekly pass, monthly LINK pass, semester pass, or whatever fits your need).
- Be aware of your surroundings.
- Keep your belongings on you at all times.

### Need a taxi?

- Metrocab Boston: 617-536-5000
- Wonderful Boston: 617-782-5500



Get a CharlieCard and Save!  
Then upgrade your CharlieCard with [MyCharlie](#)

<b>CharlieCard</b>	<b>\$2.00</b> FREE transfer to Local Bus DISCOUNTED transfer to Express Bus One Transfer valid within two hours of paying our fare.
<b>CharlieTicket/ Cash-on-board</b>	<b>\$2.50</b>
<b><a href="#">Monthly LinkPass</a></b>	<b>\$70/month</b> Valid on Subway plus Local Bus.
<b>Day/Week LinkPass</b>	<b>\$11.00</b> for 1 day <b>\$18.00</b> for 7 days Valid on Subway, Local Bus, <a href="#">Commuter Rail Zone</a> 1A, and Inner-Harbor Ferry.
<b><a href="#">Seniors and Persons with Disabilities</a></b> (Blind persons ride for free)	<b>\$1.00/ride</b> <b>\$28/month</b> Valid on Local Bus and Subway. Requires a <a href="#">Senior/T.A.P ID</a> or Blind Access Card.
<b><a href="#">Students</a></b> (Junior High and High School)	<b>\$1.00/ride</b> <b>\$28/month</b> for 7-day validity <b>\$25/month</b> for 5-day (Mon-Fri) only Valid on Bus, Subway, Express Bus, and <a href="#">Commuter Rail Zones</a> 1, 1A and 2. <a href="#">Requires Student ID Badge.</a>
<b><a href="#">Children 11 and under</a></b>	<b>Free</b> Children 11 (eleven) years of age and under ride for free when accompanied by an adult, with a limit of two children for each adult.

Subway Map



## Navigating BMC and the Health Care System

### Keeping track of medical appointments

- When you make a medical appointment make sure you get a card with the date, time and location on it.
- Write your appointment down in your calendar, or put it in your phone right away.
- You should get a reminder call and receive a reminder letter ahead of time. However, do not depend on this! Make sure you are keeping track of your own appointments.
- If you need help with transportation, call ahead of time to set up a ride, or talk with your social worker.
- If you are not going to be able to keep your appointment, call as soon as possible to reschedule – this way we will have openings to see other patients.

### Getting prescriptions filled/refilled

- Do not wait until you run out of medicine to call and ask for a refill.
- When you are seen for a clinic visit ask for any prescription refills you need.
- Prescriptions for opioid pain medication must be given to you in person; always make sure you have some pain medication at home in case you need it. When you are starting to get low, call for a refill, or ask at your next visit.

### Utilizing the Emergency Room

- If you are having pain at home page the on call doctor (617 638 5795 then pager # 5731 for the pediatric hematologist on call) for advice.
- Even if you think you need to come to the ER, paging the doctor ahead of time can help speed things along.

### Primary Care

- Your hematologist is a specialist in sickle cell disease; however there is a lot more to you than just sickle cell! It's important to have a primary care doctor who can help with any routine medical needs and also help coordinate care with different specialists. You see your primary care doctor for a regular check-up once or twice a year.

## Sickle Cell Resource List

### American Red Cross

Website: [www.newenglandblood.org](http://www.newenglandblood.org)

Telephone: 1-800-GIVE-LIFE (1-800-448-3543)

### Boston Center of Excellence in Sickle Cell Disease

Telephone: (617) 534-5727

### Department of Public Health Community Support and Care Coordination Programs for Children with Special Health Care Needs

Telephone: (800) 882-1435

### Greater Boston Sickle Cell Disease Association

Telephone: (617) 427-4100

1542 Tremont Street, Roxbury, MA 02120

<http://www.gbscda.org>

### Massachusetts Department of Public Health - Genetics Program

Telephone: (617) 534-5121

### Massachusetts Rehabilitation Commission

Website: <http://www.mass.gov/eohhs/gov/departments/mrc/>

The mission of MRC is to promote equality, empowerment and independence of individuals with disabilities.

### National Health Care Transition Center

<http://www.gottransition.org>

### National Institute of Health – Sickle Cell Anemia

Website: <http://health.nih.gov/topic/SickleCellAnemia>

### New England Pediatric Sickle Cell Consortium

Website: [www.nepscc.org](http://www.nepscc.org)

### Sickle Cell Disease Association of America

Website: [www.sicklecelldisease.org](http://www.sicklecelldisease.org)

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

### Sickle Cell Information Center

Website: [www.SCInfo.org](http://www.SCInfo.org)

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

**The Community Sickle Cell Support Group**

Telephone: (617) 427-4100

**Financial Assistance****Mass Health**

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

1-800-332-5545 (to apply for assistance)

Website: [www.hcfa.gov/medicaid/medicaid.htm](http://www.hcfa.gov/medicaid/medicaid.htm)

**Prescription for Public Transportation (PT-1 form)**

Telephone: 1-800-841-2900

**Education****Federation for Children with Special Needs**

1135 Tremont Street, Suite 420, Boston, MA 02102

Telephone: (617)-572-2094/1-800 331-0688

Website: [www.fcsn.org](http://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.

**Mass. Association of Special Education Parent**

Advisory Councils (MASSPAC)

617-962-4558

P.O. Box 167 Sharon, MA 02067

**Massachusetts Department of Education (DOE)**

(781) 338-3000

Bureau of Special Education Appeals

(781) 338-6400

Program Quality Assurance

(781) 338-3700

350 Main Street

Malden, MA 02148

[www.doe.mass.edu/sped](http://www.doe.mass.edu/sped)

**Sickle Cell Disease: Information for School Personnel**

Website: [www.state.nj.us/health/fhs/sicklecell](http://www.state.nj.us/health/fhs/sicklecell)

**Special Kids Special Help – Boston Medical Center**

Web based resource for parents with children with special needs.

Website: [www.specialekidsspecialhelp.org](http://www.specialekidsspecialhelp.org)

**Camps and Programs for Teens with Sickle Cell Disease****Boston Medical Center STRIVE**

Telephone: 480-518-3216 (Megan Garland) or 207-523-0822 (Victoria Hoch)

Offers Mentoring, Tutoring and Advocacy for Teenagers with Sickle Cell Disease.

**Camp Sunshine**

Telephone: (207)655-32800

Website: [www.campsunshine.org](http://www.campsunshine.org)

Located in Casco, Maine. Offers free sessions for families. Special sessions available for specific illnesses.

**The Hole in the Wall Gang Camp**

Telephone: (860) 429-3444

Website: [www.holeinthewallgang.org](http://www.holeinthewallgang.org)

Located in Ashford, Connecticut, this camp offers free sessions for children age 7 to 15. Special sessions are available for specific illnesses

**Legal Aid****Disability Law Center**

(800) 872-9992

11 Beacon Street, Suite 925(800) 872-9992

Boston, MA 02108

**Massachusetts Advocacy Center**

(617) 357-8431

100 Boylston Street, Suite 200

Boston, MA 02116

**Parent Professional Advocacy League (PAL)**

(617) 227-2925

15 Court Street, Suite 1060

Boston, MA 02108

[www.ppal.net](http://www.ppal.net)

**Patient Advocate Foundation**

Website: [www.patientadvocate.org](http://www.patientadvocate.org)

Telephone: 1-800-532-5274

Provides education and legal counseling on managed care, insurance, and financial issues for Sickle Cell patients

**Volunteer Lawyers Project**

(617) 423-0648  
(VLP of the Boston Bar Association)  
29 Temple Place, 3rd Floor, Boston, MA 02111

**Government Agencies****Health Care for All**

Health Care for all is a Massachusetts organization dedicated to making adequate and affordable health care accessible to everyone, regardless of income, social or economic status. Their website has detailed information on health insurance programs: [www.hcfama.org](http://www.hcfama.org). They have a free helpline service and can be reached at 1-800-272-4232.

**Massresources.org**

This is a free online resource for Massachusetts residents in need of housing, food, health care, and other basic services. [www.massresources.org](http://www.massresources.org)

**Massachusetts Department of Mental Health**

(800) 221-0053  
25 Staniford Street  
Boston, MA 02114

**Massachusetts Department of Public Health**

(617) 624-6000  
250 Washington Street  
Boston, MA 02108

**Massachusetts Department of Social Services**

(617) 748-2000  
24 Farnsworth Street  
Boston, MA 02108

**Massachusetts Rehabilitation Commission**

1-617-204-3603  
[www.mass.gov/eohhs/gov/departments/mrc/](http://www.mass.gov/eohhs/gov/departments/mrc/)

**U.S. Office for Civil Rights**

(617) 223- 9662  
FAX (617) 223-9669  
U.S. Department of Education  
J.W. McCormack P.O.C.H., Room 707  
Boston, MA 02109  
(for section 504 and ADA complaints)



<p><b>My Sickle Cell History</b></p> <p>Type:</p> <p>Current Medications:</p> <p>Complications:</p> <p>Baseline labs:</p> <p>Allergies:</p>	<p><b>Contact Information</b></p> <p>BMC Emergency Contact:</p> <p>My Hematologist:</p> <p>My Primary Care Provider:</p> <p>Other Providers:</p>
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## My Sickle Cell Disease

I have Sickle Cell Disease. The type I have is:

HbSS \_\_\_\_\_

HbSC \_\_\_\_\_

HbS Beta Plus Thalassemia \_\_\_\_\_

HbS Beta Zero Thalassemia \_\_\_\_\_

Other \_\_\_\_\_

2. My Baseline Hemoglobin is \_\_\_\_\_

White count is \_\_\_\_\_

O<sub>2</sub> Saturation is \_\_\_\_\_

3. In the past I have experienced:

- Acute Chest Syndrome YES \_\_\_\_\_ NO \_\_\_\_\_

- Gallbladder Disease YES \_\_\_\_\_ NO \_\_\_\_\_

- Splenic Sequestration YES \_\_\_\_\_ NO \_\_\_\_\_

- A Cholecystectomy YES \_\_\_\_\_ NO \_\_\_\_\_

- A Splenectomy YES \_\_\_\_\_ NO \_\_\_\_\_

4. My Primary Care Doctor is: \_\_\_\_\_ Phone # \_\_\_\_\_

My Hematologists are: \_\_\_\_\_ Phone # \_\_\_\_\_

\_\_\_\_\_ Phone # \_\_\_\_\_

My Cardiologist is: \_\_\_\_\_ Phone # \_\_\_\_\_

My Neurologist is: \_\_\_\_\_ Phone # \_\_\_\_\_

My Pulmonologist is: \_\_\_\_\_ Phone # \_\_\_\_\_

My Ophthalmologist is: \_\_\_\_\_ Phone # \_\_\_\_\_