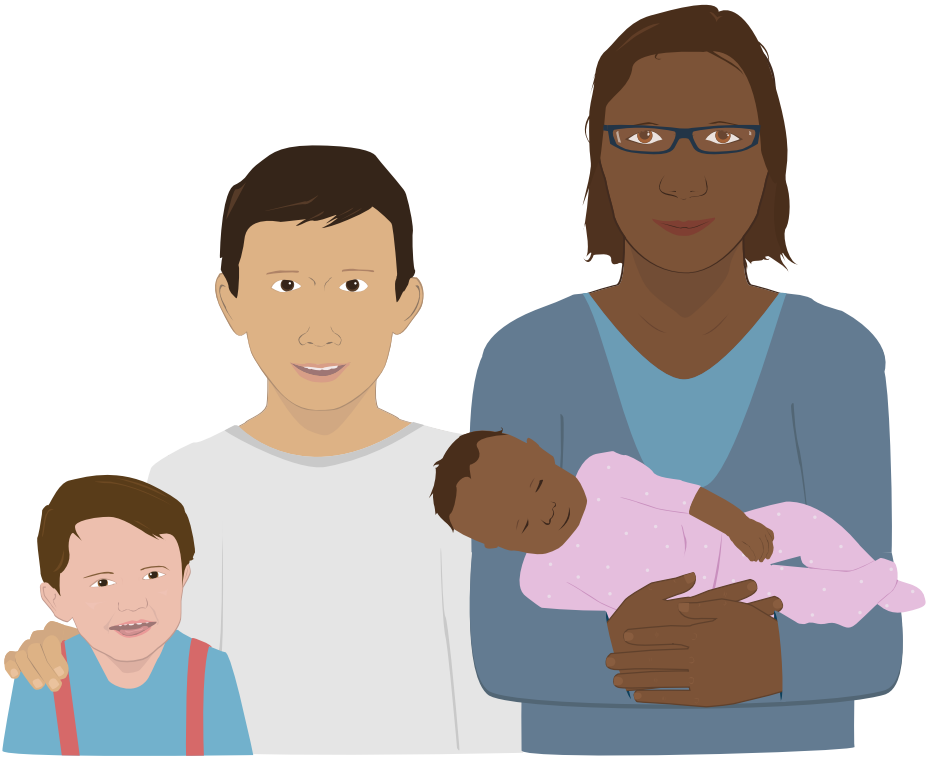
Sickle Cell Disease

A Family Handbook

This handbook is for:

.....

This handbook is to help you and your family understand sickle cell disease and manage your child's care.



Centre universitaire de santé McGill

McGill University Health Centre

Québec

This document was developed by the following:

John Kayser, RN, MSc.
Marie Gale, RN, BSc.
Anelise Santo, RN, MSc.
Sharon Abish, MD.
Debbie Pealow, SW, MSc.
Elissa Remmer, RN, MSc.
Julie Brouillard, RN, MSc.
Josephina Revulta, RN, BSc.
Christina Rosmus, RN, MSc.

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IMPORTANT: PLEASE READ

This handbook is for information only. It is not to take the place of the advice you receive from your healthcare professional. As every patient is different, different treatments are chosen for each patient. Speak to a member of your healthcare team if you have questions or concerns.

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Office d'éducation des patients du CUSM
MUHC Patient Education Office

MM
Projet d'informatique médicale Molson de McGill
McGill Molson Medical Informatics

This material is also available on:
MUHC Education Collection (http://infotheque.muhc.ca/),
MUHC Patient Education Office (www.muhcpatienteducation.ca) and
Ministry of Health and Social Services: www.msss.gouv.qc.ca/depistage-neonatal

Table of contents

Look out for sections with **Information Sheets** indicated by this symbol!

Dear parents,

Having a child newly diagnosed with sickle cell disease is a stressful time for most families — you are not alone. Other parents are living the same experience as you and there is a health team here to support you. You probably have many questions on your mind such as:

"How will this illness affect my child's life now and in the future?"

"What are the symptoms of this disease, what do they mean and what do I do about them?"

"Will my child be able to lead a normal life?"

These are some of the questions that drive many parents to learn more about sickle cell disease so that they can provide the best possible care to their child.

Based on our experience and from what parents have told us, we have designed this handbook so that it is **useful, practical and easy to read.**

It will provide you with valuable information about sickle cell disease that can help you answer many of the questions you may have and hopefully help you adapt to this stressful situation.

The hematology health team

How to Use This Handbook

Many parents feel overwhelmed by the amount of information given to them about sickle cell disease. They worry that they will not be able to remember everything that is being said about their child's health. This is quite normal. To facilitate your reading and understanding, this handbook is divided into three parts and contains detailed Information Sheets.

Part 1: Signs and symptoms: what to look for
The first part contains information that will be very useful to you at this point. It includes a list of symptoms to look for, when and who to call for advice and when to get medical attention.

Part 2: Understanding Sickle Cell Disease
In the second part, you will find more detailed explanations about sickle cell disease, and how it is passed on.

Part 3: Recognizing potential complications and how to help prevent them
The third part of this handbook includes information about possible medical crises and complications that your child may experience due to sickle cell disease.

Keep in mind that not every child with sickle cell disease will experience these difficulties. Most children affected by the illness can lead happy and fulfilled lives.

We encourage you to read this handbook and discuss it with your doctor, nurse or other members of your treating team at the hospital. Do not hesitate to ask any unanswered questions you may have.

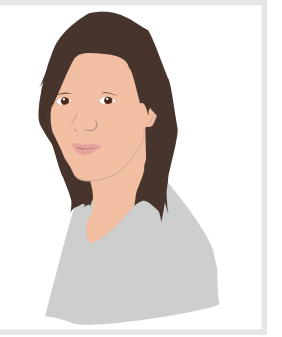
Introduction

In Québec, there are 4 centres who specialize in the diagnosis and treatment of children with sickle cell disease and other forms of severe hemoglobinopathies:

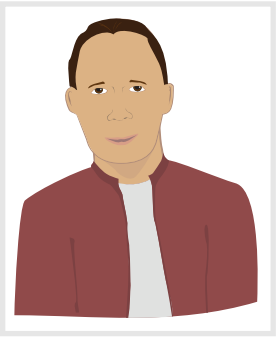
- Centre Hospitalier Universitaire de Québec - Centre Hospitalier de l'Université Laval
- Sherbrooke University Hospital Centre - Fleurimont Hospital
- McGill University Health Centre - Montreal Children's Hospital
- Sainte-Justine University Hospital Centre

Questions or Concerns — How to Get Answers?

The **hematologist** is a doctor who specializes in diseases of the blood such as sickle cell disease. When you come to the hospital you will meet many doctors. All of these doctors are there to help you and your child. Besides your regular appointments with the hematologist, we strongly encourage you to keep taking your child to the pediatrician or to your family doctor because you will still need medical care that does not require expertise from a hematologist.

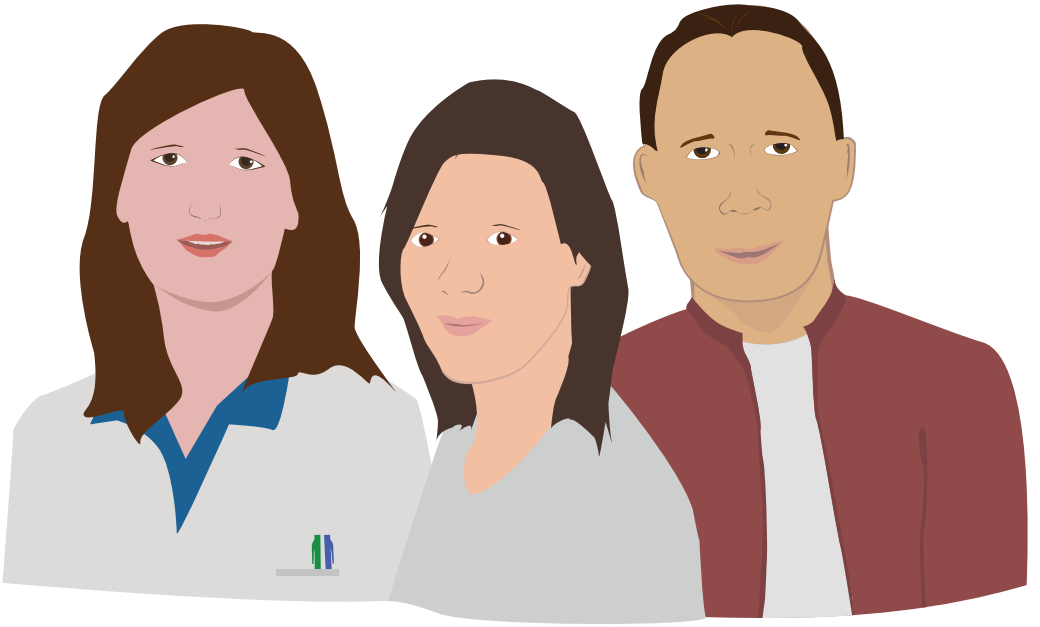


The **sickle cell nurse** clinician is a helpful person to contact to ask your questions about your child's health. She will meet with you at diagnosis and offer you information that will be very helpful. Along with the other healthcare team members, she will help you reach a better understanding of your child's illness, and may also help you manage some of the symptoms. If your child needs to see a doctor, the sickle cell nurse clinician may facilitate your visit to the clinic or to the emergency room depending on your child's condition.



Social Services

Social workers assist with concrete needs: meals, transportation, financial issues, supportive counseling, school issues, crisis intervention, compliance issues, and ethical dilemmas.
They will assess and evaluate each situation and assist with the appropriate resources.



The hematology team consists of several healthcare professionals that will work with you to help you and your child manage this new life situation. You will find a list of these professionals on page 41 of this guide.

Part 1: Signs and symptoms: what to look for

When Does My Child Need Medical Care?

Sickle cell disease is a medical condition that can have potential serious consequences if some symptoms are left untreated. Seeking treatment after early recognition of symptoms is very important. Contact us if you are concerned about your child's health. The information sheet called "Hematology team" has all the important telephone numbers...always keep it handy!

Signs and symptoms:

- fever
- prolonged pain
- distended or enlarged abdomen
- difficulty breathing
- painful and persistent penile erection
- headache
- stiff neck
- weakness or numbness of arms or legs
- change in vision
- severe abdominal pain

If your child experiences any of these symptoms, you must see a doctor. Call the hematology team and take your child to the hematology clinic or to the emergency room.

Description of symptoms. Contact us and see a doctor if...

Do not forget to inform the emergency team that your child has sickle cell disease.

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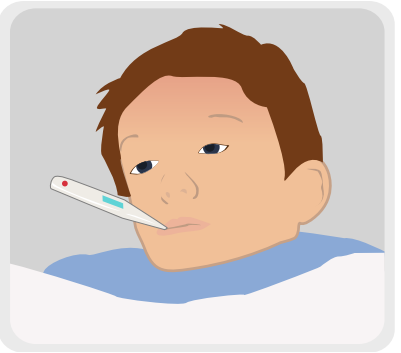

Fever

...your child has a temperature greater than:

- 38.5° C (101° F) rectally, or
- 38° C (100.4° F) orally, or
- 37.5° C (99.3° F) axillary. A fever must not be ignored in a child with sickle cell disease.

Report fever to your doctor or nurse and take your child to the hematology clinic or to the emergency room immediately. Do not wait.

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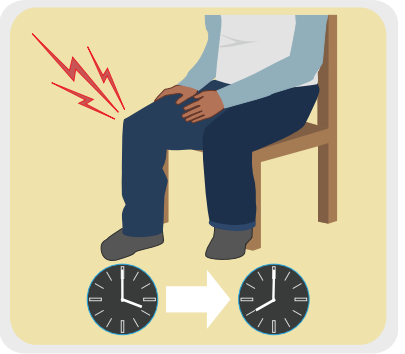
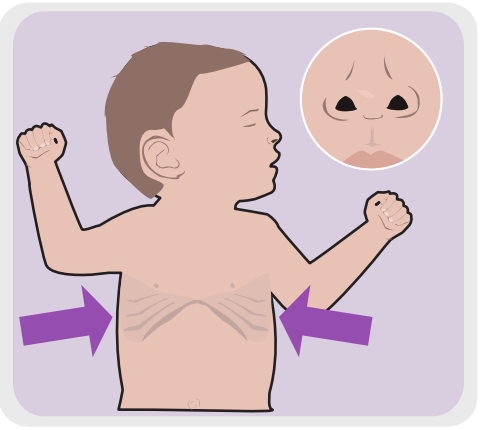


Prolonged pain

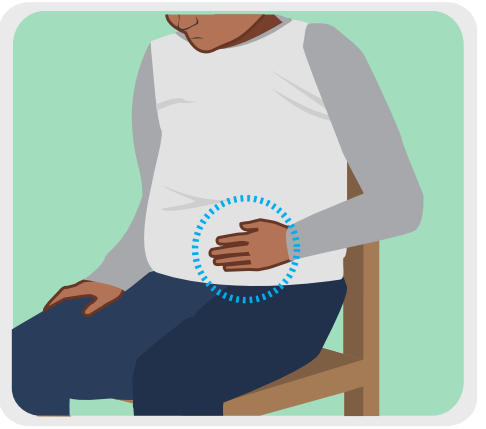
...your child is feeling pain that is not relieved by rest, increased fluids, acetaminophen (ex. Tylenol™ or Tempra™), ibuprofen (ex. Advil™) or morphine/codeine if it is prescribed by your physician. Look for the information sheet called “Pain management” for more details.



Difficulty Breathing

…your child experiences one or more of these symptoms:

- rapid breathing
- shortness of breath
- persistent cough
- chest pain



Distended or Enlarged Abdomen

…your child's belly becomes enlarged and feels hard to the touch. This might mean that the blood cells are getting trapped in the spleen, causing it to enlarge. Your doctor will teach you how to feel your child's spleen.

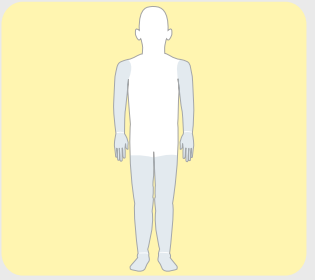
Painful and persistent penile erection

…your child (boy) develops a painful, penile erection that lasts more than 1 hour.

You should also contact us and bring your child to the hematology clinic or to the emergency room if you notice any of the following symptoms in your child:

Headache

Stiff Neck



Weakness or Numbness of Arms or Legs

Change in Vision

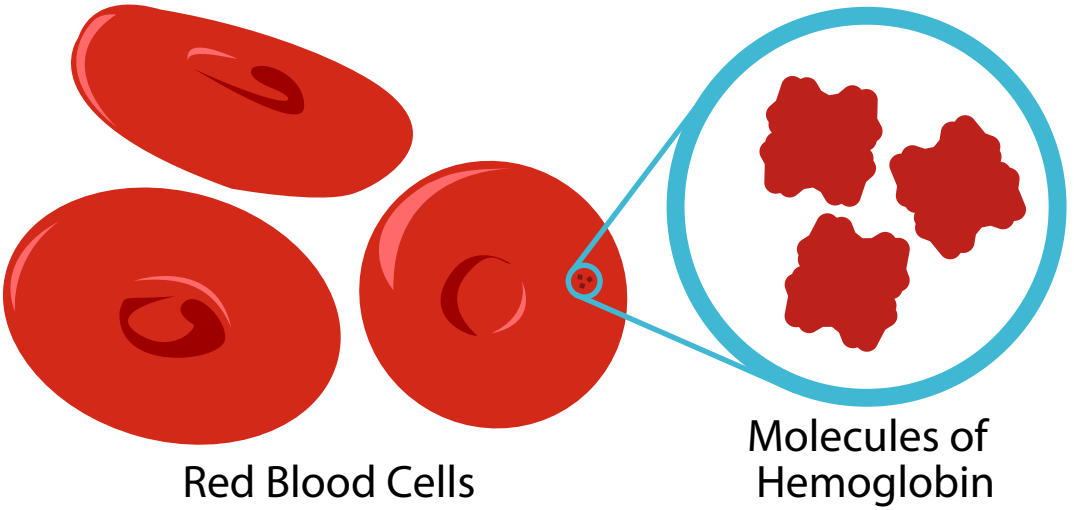
Severe Abdominal Pain

Do not forget to inform the emergency team that your child has sickle cell disease.

Part 2: Understanding Sickle Cell Disease

What is Sickle Cell Disease?

Sickle cell disease is an illness that affects the hemoglobin that is contained in your child's red blood cells. **It is often diagnosed during early childhood.**

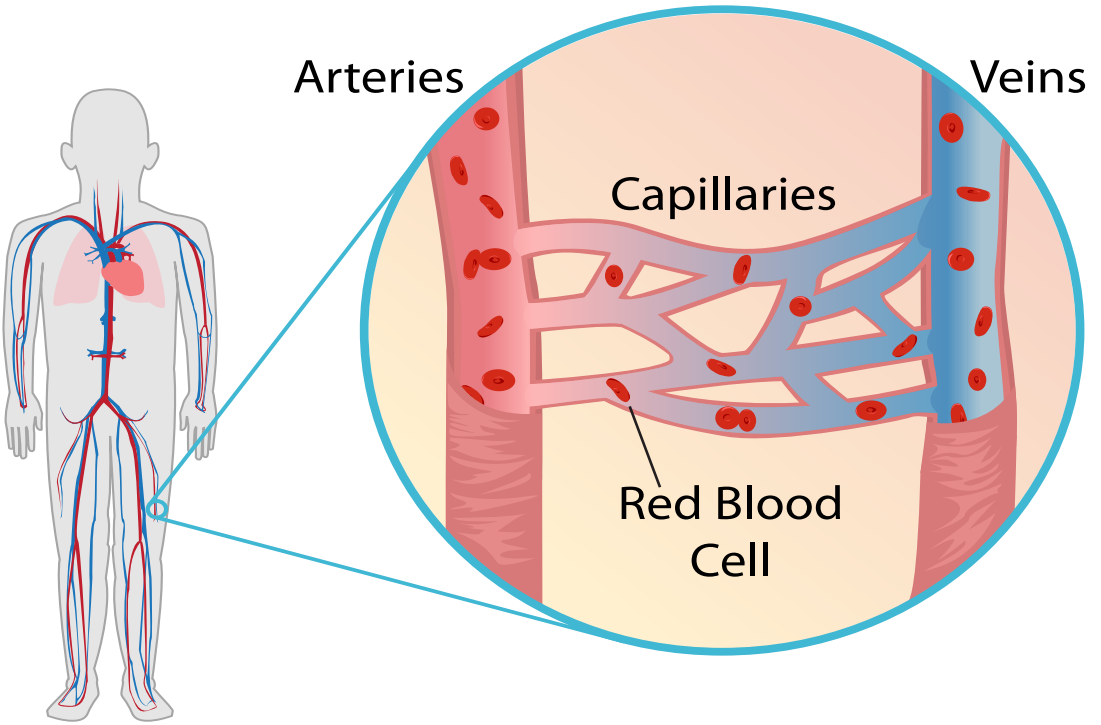


- It is a chronic illness, but different treatments and medications can help in managing some of the symptoms and complications. For a better understanding of the disease, it is important to understand how blood normally flows through the body.

How does blood normally flow through the body?

Blood flows through blood vessels by a complicated system of arteries, veins and capillaries.

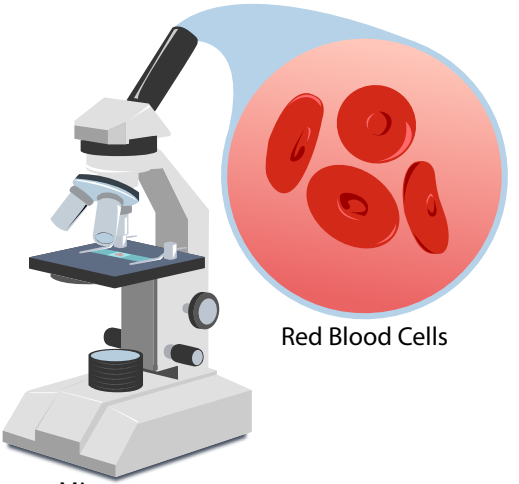
- **Arteries** are elastic-like tubes that carry the blood from the heart to the rest of the body (for example, your brain, eyes, liver, and muscles).
- **Veins** carry the blood from the body parts back to the heart.
- **Capillaries** are the tiny elastic-like tubes that connect the arteries to the veins. A capillary is about 1000 times smaller than a vein or an artery.



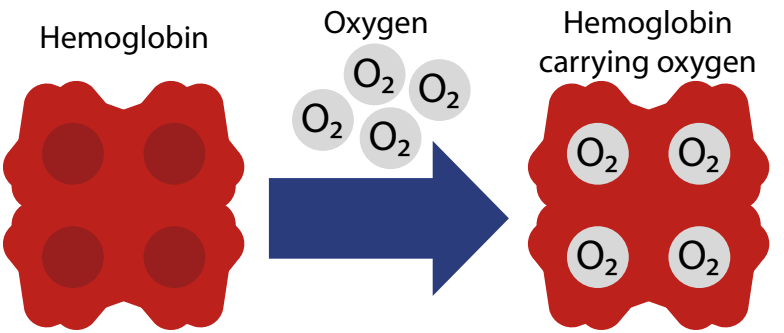
The **vascular system** is made up of arteries, veins and capillaries

Part 2: Understanding Sickle Cell Disease

Blood is mostly composed of microscopic cells called red blood cells. Normal red blood cells are disk-shaped and flexible. Their shape and flexibility allows them to flow freely through the arteries and veins and 'squeeze' through the very narrow passages of the capillaries.



Each red blood cell is filled with small molecules called hemoglobin. An important role of hemoglobin is to transport the oxygen to the different parts of the body. This is important because our body depends on oxygen. Oxygen is like gas for our car — if there is little or no gas in our car, the car will not work properly.

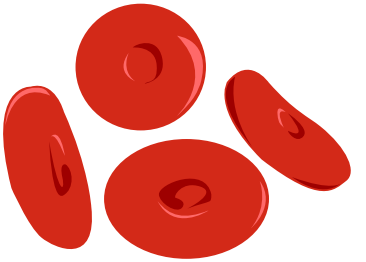


Hemoglobin picks up oxygen and delivers it to other parts of the body.

How does the blood flow differently when you have sickle cell disease?

In children with sickle cell disease, many of the red blood cells are “sickle-shaped”. In other words, many of the cells are in the form of a crescent. Furthermore, sickle cells are stiff and less flexible than regular red blood cells. As explained on the previous page, normal red blood cells can squeeze through the very narrow capillaries because they are disk-shaped and flexible. Sickle-shaped red blood cells do not pass through so easily. This means that many red blood cells may get stuck in the capillaries creating a blockage.

Sickle Shaped
Red Blood Cells



Normal
Red Blood Cells

Sickle cells are **stiff** and **not very flexible**. This allows them to create blockages in the blood vessels.

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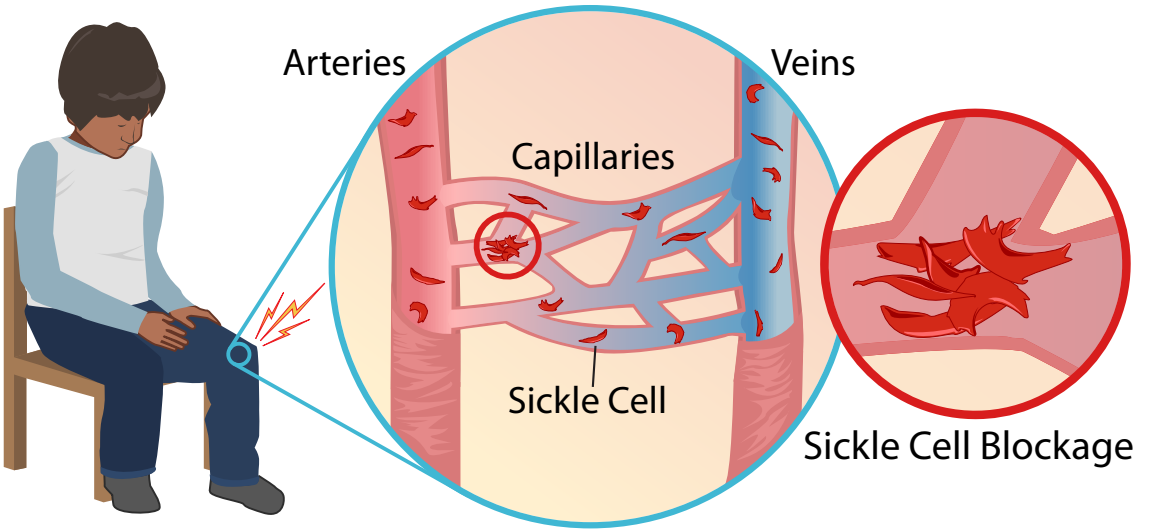
Because the blood flow is decreased by this blockage, your child may experience a variety of symptoms caused by a lack of blood and oxygen to some parts of the body. This is the cause of most “sickle cell crises.” Pain is one of the most common symptoms of a sickle cell crisis. It can range from mild to severe and varies in intensity, location and duration.



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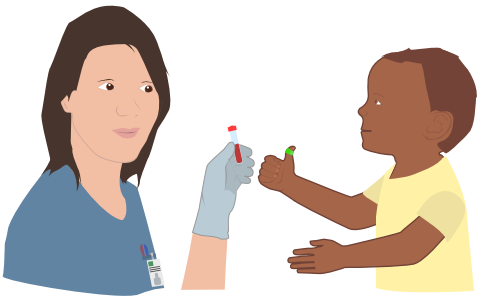
Many factors can contribute to these events. Some common examples are fever, dehydration, exposure to cold weather, sudden weather changes, vigorous exercise and stress. However, most often these crises happen without any specific reason. They are unpredictable and can happen slowly or suddenly.

Why do most children with sickle cell disease have a low hemoglobin?

Sickle cells have a shorter life-span than normal red blood cells. They are destroyed earlier because of their odd shape. For example, normal red blood cells live about 120 days while sickle cells live for approximately 20 days.

The body constantly makes new red blood cells but in this case, it cannot make the cells as fast as they are destroyed. So a child with sickle cell disease, usually has less red blood cells in the body. Fewer red blood cells also mean less hemoglobin, therefore less oxygen.



A blood test measures your child's hemoglobin level.

Here are some symptoms of low hemoglobin:

- **Your child is more tired**
- **Your child's eyes become yellow**

If you notice these symptoms, call us. A hemoglobin check may be necessary.

Your child will need to have a blood test to measure the hemoglobin level at each clinic visit. This level will vary from time to time depending on your child's condition. A lower than usual hemoglobin level might require more frequent medical visits in order to monitor the changes in that level. Sometimes, a blood transfusion may be necessary.

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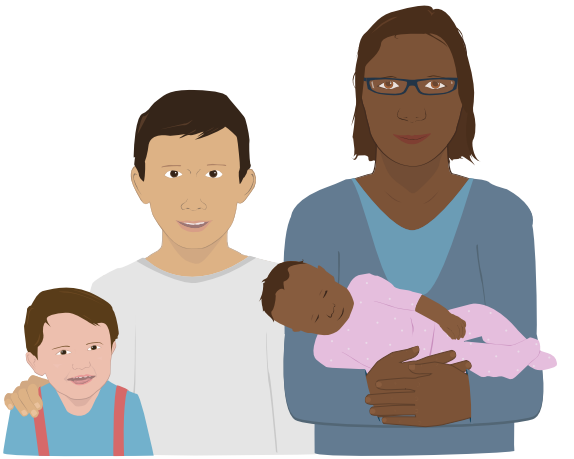
To help you keep track of your child's hemoglobin, we have included an information sheet called, "Complete Blood Count Log." **Please bring this sheet to your clinic visits and record your child's hemoglobin levels.**

How Did My Child Get Sickle Cell Disease?

Sickle cell disease is **a genetic disorder**. It is inherited, which means that your child was born with it. **It is not contagious.**

How did the sickle cell genes get passed on to my child?

Each child gets two hemoglobin genes, one from the mother and one from the father. Genes are the body's substances found in each cell that determine everything about us, from the color of our eyes, skin, hair and also our hemoglobin type.



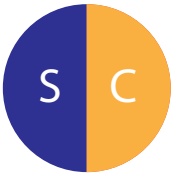
In order to have sickle cell disease, a child needs to inherit one sickle cell hemoglobin gene (HbS) from one parent and one abnormal hemoglobin gene (ex. HbC) from the other parent.

What type of sickle cell disease does my child have?

There are three common types of sickle cell disease:

1. Sickle Cell Anemia (Hemoglobin SS Disease or HbSS)

- This is the most common type of sickle cell disease
- Children with sickle cell anemia have almost 100% hemoglobin S in their red blood cells
- They inherit a hemoglobin S gene from each parent



2. Sickle Cell Hemoglobin C Disease (Hemoglobin SC Disease or HbSC)

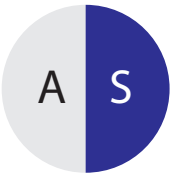
- Children with sickle cell hemoglobin C disease inherit a hemoglobin S gene from one parent and a hemoglobin C gene from the other parent
- Hemoglobin C is another type of abnormal hemoglobin

3. Sickle Cell Beta-Thalassemia Disease (hemoglobin SBthal or HbSβThal)

- Children with sickle cell beta-thalassemia inherit a hemoglobin S gene from one parent and a beta-thalassemia gene from the other parent
- Beta-thalassemia is another type of abnormal hemoglobin
- These children have mostly hemoglobin S in their red blood cells

Sickle cell carrier or sickle cell trait

If a child gets one sickle cell hemoglobin gene (HbS) from one parent and receives a normal hemoglobin gene (HbA) from the other, the child will have sickle cell trait or be a sickle cell trait carrier. A person with sickle cell trait does not have sickle cell disease but carries the sickle cell gene. People with sickle cell trait are **usually** healthy**.** They do not have symptoms of the disease. They may however, pass their sickle cell gene on to their children.



Hemoglobin S Carrier

- Sickle cell trait
- This is **not** a form of sickle cell disease

There is no proven link between being a carrier of the gene and the possibility of developing symptoms of the disease. Although in some extreme conditions (ex. scuba diving, low oxygen in the air, mountain climbing at high altitudes, very strenuous exercise and severe dehydration), rare cases of health problems among carriers have been reported.

Individuals with sickle cell trait should receive informative genetic counseling. This will help them know their chances of having children with sickle cell disease.

> **Because the disease is so unpredictable, it is difficult to know which children will have more symptoms than others.**

As indicated on the previous pages, all three types of sickle cell disease are characterized by the presence of hemoglobin S (HbS) in the red blood cells but the amount will vary from one type to the other. It is HbS that will cause the red blood cells to become rigid, elongated and sickle shaped.

Based on the literature and from our experience, most children with sickle cell hemoglobin C disease and some children with sickle cell beta-thalassemia often have milder symptoms.

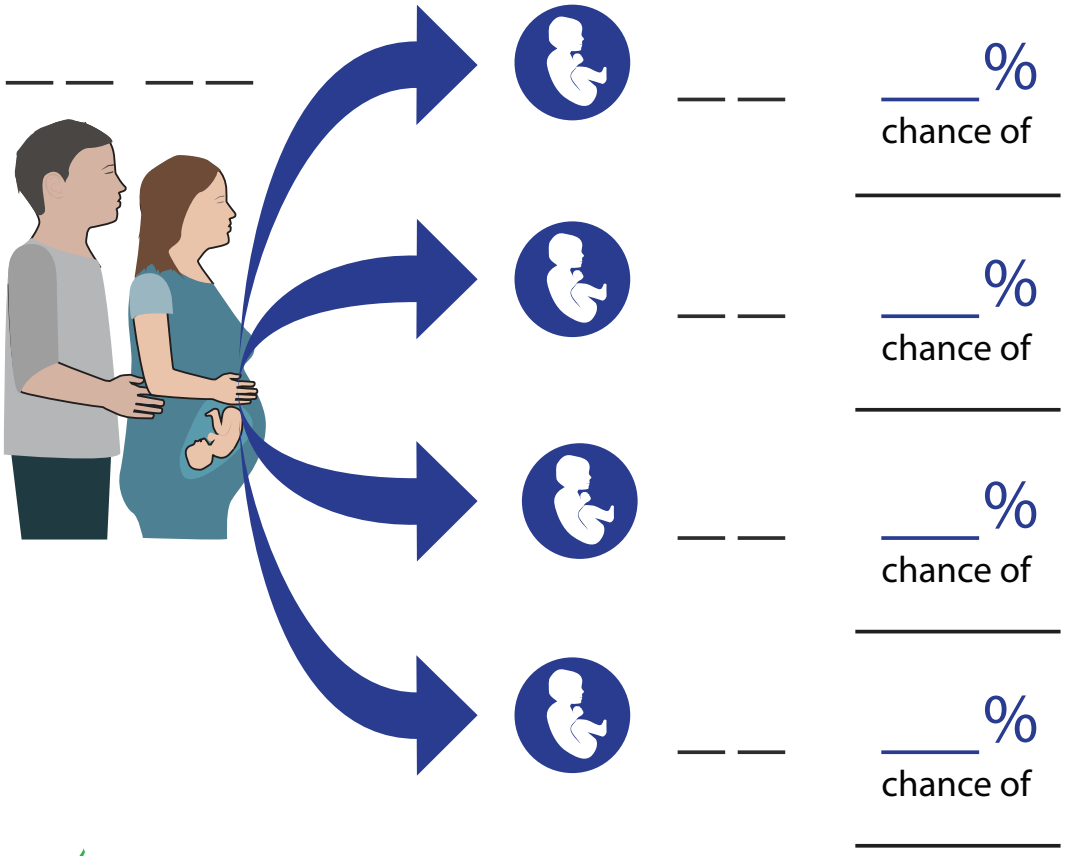
Your child's sickle cell disease type was determined through a blood test. This blood test identifies the different types of hemoglobin and measures the amount (%) of HbS in his red blood cells.

Generally children with sickle cell disease start to experience symptoms during the first year of life, usually after six months of age. Symptoms and complications may be different for each child and may range from mild to severe. It is important to note that there is no single best treatment for all children with sickle cell disease.

> **Treatment decisions will depend on the child's condition and on the symptoms.**

Part 2: Understanding Sickle Cell Disease

A staff member will help you complete this diagram so you can see how sickle cell disease might occur in your family with each pregnancy.



Please **check-off** your child's sickle cell disease type:

- ☐ Sickle Cell Anemia (HbSS)
- ☐ Sickle Cell Hemoglobin C Disease (HbSC)
- ☐ Sickle Cell Beta-Thalassemia (HbSβThal)

> Sickle cell disease is one of the **most common** genetic disorders.

Are you planning to have another child?

If you are pregnant or planning on having other children, genetic counseling is available to you at all four designated centres. Genetic counselors are experts in hereditary disorders. They will help you figure out the likelihood of having future children with sickle cell disease. They can also offer you options for family planning and prenatal testing.

Cord blood collection from your future new born baby can also be a very interesting option that may benefit your child with sickle cell disease. If your child is a candidate and the cord blood is compatible, a stem cell transplant could be indicated. We encourage you to discuss these options with your doctor or your nurse. Please let us know if you are pregnant or planning to have other children.

Who gets sickle cell disease?

Sickle cell disease affects millions of people around the world. In the United States, about 1,000 children are born each year with the disease. This makes it one of the most common genetic disorders in the United States. Out of every 375 births of African American parents, one child is born with sickle cell disease. It happens most often to children whose parents are from sub-Saharan Africa, southern India, the Mediterranean, South America and the Caribbean.

In Québec, about 475 children between 0 to 18 years old are effected with sickle cell disease and are being followed in the four specialized centers.

Part 3: Recognizing potential complications and how to help prevent them

You will find that a **better understanding** of the possible health problems that may happen in sickle cell disease will help you be prepared and gain **more control** of your child's health.

In the 3rd part of this handbook, we cover the most common symptoms along with some of the potentially serious complications of the illness. Take your time to read this information and please ask us any questions that you may have.

Two of the most common health concerns in children with sickle cell disease are **infection** and **pain.** It is important to know what to look for so you can help your child when these uncomfortable symptoms occur.

Infection

Fever may be a sign of a potential infection. It is a symptom that should be taken seriously in children with sickle cell disease. Fever is common in children and will occur for many reasons. Children with sickle cell disease are at higher risk of getting serious infections because their spleen has been damaged at a very young age (about 4 months) by the sickle cells.

The spleen is the organ of the body that filters and removes bacteria from the blood stream. The damaged spleen of sickle cell children will not filter properly. This may allow bacteria into the blood stream. This potentially serious medical condition is called sepsis and may be very serious and even fatal. Adequate vaccination, penicillin or an alternative antibiotic taken daily as ordered by the doctor are efficient and offer good protection against infections.

Call us if possible and take your child to the hematology clinic or to the emergency room if your child's temperature is:

- greater than 38.5° C (101° F) rectally
- greater than 38° C (100.4° F) orally (in the mouth)
- greater than 37.5° C (99.3° F) axillary (under the arm)

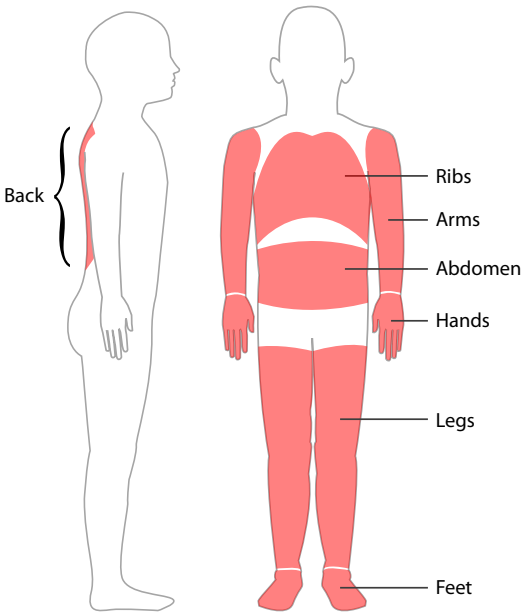
Remember: Do not ignore a fever in a child with sickle cell disease! See the information sheet: "The Hematology Team" for important telephone numbers.

Other potentially serious infections include:

- **Meningitis** (infection in the spinal cord and brain)
- **Pneumonia** (infection in the lungs)
- **Osteomyelitis** (infection in the bones)

Pain

Pain is another symptom that is common in children with sickle cell disease. During a sickle cell crisis, sickle cells create a blockage in the blood vessels resulting in a decrease of blood flow and oxygen to certain body parts causing pain.



The best way to manage the pain is to treat it as soon as it starts. Pain crises may vary in terms of location, intensity, and frequency and can range from mild to severe. Most of them may be treated at home but in some situations, hospitalization may be required.

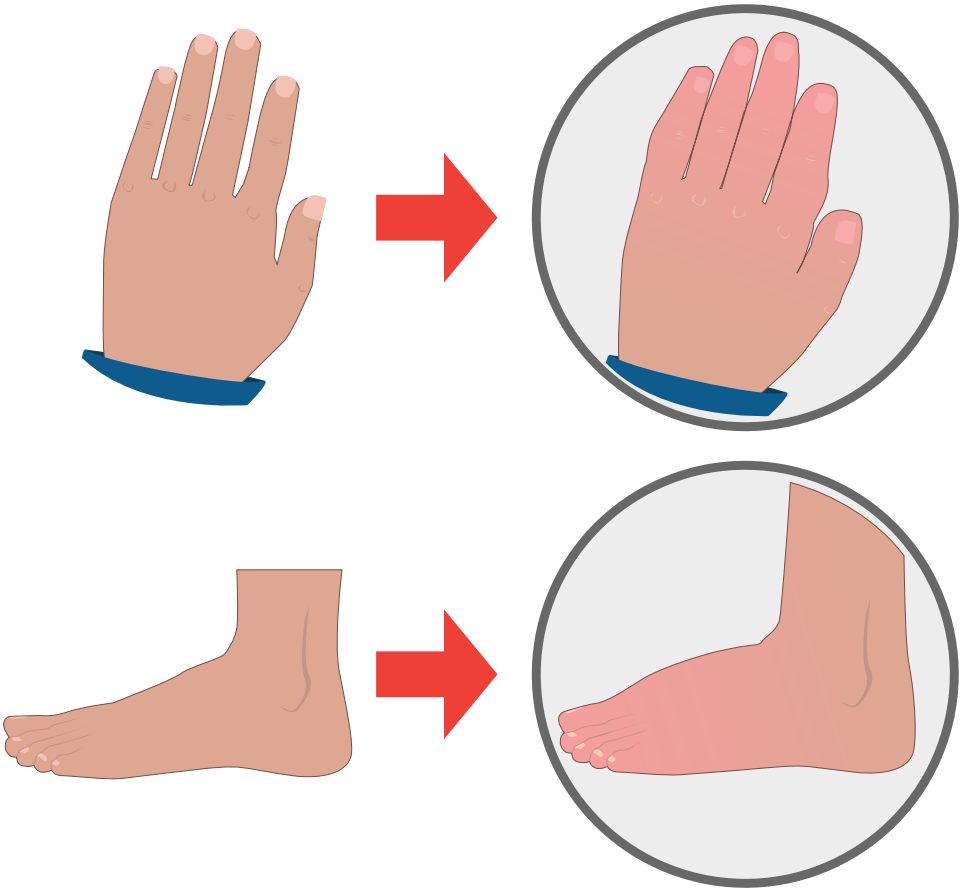
Should your child experience frequent pain episodes, the hematology team, along with pain services, can help you find the best way to manage your child's pain.

During a sickle cell crisis, your child may experience pain in the following areas: back, ribs, arms, abdomen, hands, legs or feet.

Tips for knowing if your child is in pain:

- Crying inconsolably
- Clutching the painful area
- Refusing to use a leg or arm
- Refusing to walk
- Swelling in the painful area

Note: In young children (usually under 2 years), a sickle cell crisis can cause swelling of hands or feet (fingers and toes). This is called dactylitis. This condition may last a few days to 1 week and may be accompanied with fever.



Other Possible Health Problems

Children with sickle cell disease may experience a variety of health problems that can be potentially serious. Being aware of these conditions, early recognition of the symptoms, along with quick medical intervention, are essential for optimal management of the situation. Here are some of the potential health problems:

- Acute Chest Syndrome
- Stroke
- Enlarged spleen (splenic sequestration)
- Prolonged and painful erection (priapism)
- Aplastic Crisis (severe anemia caused by a viral infection)
- Avascular Necrosis
- Retinopathy

Acute chest syndrome

Acute chest syndrome is a serious health problem that needs to be treated immediately. It is a common reason for hospitalization among children with sickle cell disease, and may represent a medical emergency. Acute chest syndrome happens when blood flow is blocked by sickle cells in the lungs. Pneumonia can trigger an acute chest syndrome which may also be associated with a pain crisis. These symptoms can range from very mild to severe and may change quickly.

Here are some of the symptoms associated with acute chest syndrome:

- Chest pain
- Fast breathing , or trouble breathing
- Congested cough
- Fever
- Abdominal pain

If your child experiences any of these symptoms, you must see a doctor. Call the hematology team and take your child to the hematology clinic or to the emergency room. Do not wait.

Stroke

A stroke happens when a part of the brain is deprived of blood supply and oxygen. Children with sickle cell disease are more at risk of having a stroke than other children. It is most likely to happen between the ages of 2 to 10 years old. A stroke is a sudden and severe complication of the illness that represents a medical emergency.

Here are some of the symptoms associated with a stroke:

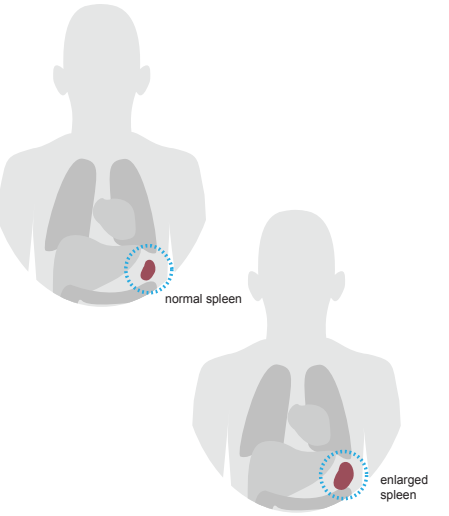
- Loss of consciousness (like rapidly falling asleep and falling to the ground)
- Weakness of arms and/or legs
- Difficulty talking or slurred speech
- Unsteady walk
- Complains of changed vision
- Unable to move legs or arms or one side of the body
- Seizures
  - Loud groaning
  - Losing consciousness
  - Body becomes rigid
  - Saliva or foam may drip from the mouth
  - Sweating, tremors, quick movements of arms and legs

If you suspect that your child is having a stroke, call 911. Your child must be brought to the emergency room right away.

7

Your doctor may order a special test for your child that may help detect narrowing in the blood vessels of the brain. This information is critical in helping us possibly prevent a stroke in the future. This test is called a Transcranial Doppler (TCD) Ultrasound. An ultrasound allows the team to view what is happening in the brain as a live video. This is a very important test. Do not miss your appointment for this test. The decision to order such a test will depend on the type of sickle cell disease, your child's age and clinical evaluation by the doctor. We encourage you to discuss this test with your doctor or nurse.

Splenic Sequestration (Enlarged Spleen)



Splenic sequestration is a condition that occurs when the blood becomes trapped in the spleen, causing it to become larger than normal. The spleen is an organ that is located on the left side of your child's abdomen.

As mentioned earlier, the spleen of children with sickle cell disease has been damaged by sickle cells from a young age. Red blood cells may become trapped in the spleen because the vessels leading in and out of it have also been damaged. When this happens, the spleen can become very large and easy to feel. This condition is called "splenic sequestration." Depending on the severity of this condition, your child's hemoglobin may decrease. Sudden enlargement of the spleen, with a large decrease in hemoglobin level is a serious condition. A blood transfusion may be necessary and it could become an emergency. If the spleen becomes larger over several weeks, and the blood hemoglobin level is stable, it is not as serious. Nevertheless, any enlargement of the spleen should be closely monitored for any changes.

Here are some of the symptoms associated with enlarged spleen

- Irritability
- Unusual weakness or feeling faint (unusual sleepiness)
- Lips and mucus membranes (inside the mouth) become very pale
- Heart beat is faster than usual
- The abdomen becomes larger in size
- Pain to the left side of the abdomen

If your child experiences any of these symptoms, you must see a doctor. Call the hematology team and take your child to the hematology clinic or to the emergency room. Do not wait.

Aplastic Crisis

An aplastic crisis happens when the body stops making new red blood cells. This condition is often caused by a virus called parvovirus B19. If this happens, your child's hemoglobin could drop to worrisome levels in a matter of days. This condition is usually treated with blood transfusions until the body starts making new red blood cells again.

Here are some symptoms associated with aplastic crises:

- Full body weakness
- Lethargy (feeling drowsy and sluggish)
- Paleness (for example, ash-like grey color in the lips and tongue. If your child has lighter color skin, the color may look more bluish.)
- Feeling dizzy or fainting
- Headache

If your child experiences any of these symptoms, you must see a doctor. Call the hematology team and take your child to the hematology clinic or to the emergency room. Do not wait.

Priapism (prolonged and painful erection)

Priapism is a serious health problem that may happen to boys living with sickle cell disease. It is characterized by a persistent and unwanted erection of the penis that can last several hours, caused by difficult drainage of the blood out of the penis vessels due to the sickle cells. This condition can be extremely painful and in some instances can lead to impotency. Priapism events may begin during sleep or during everyday activities. Episodes can last for 30 minutes to several hours and may begin in early childhood or at anytime in life. Pain management and blood transfusions may be indicated. If your child reports this condition or if you suspect this condition, talk to your doctor or nurse about it.

If this condition occurs and lasts more than 1 hour, you must see a doctor. Call the hematology team and take your child to the hematology clinic or the emergency room. Do not wait.

Avascular Necrosis

Avascular necrosis is a condition that refers to damage caused by sickle cells in the bones and joints. This most often occurs in the hips and the shoulders. It causes pain and may make walking difficult. If your child reports hip pain, please consult your doctor. Early detection and management of avascular necrosis is important.

Retinopathy

Sickle cell disease has the potential to cause eye complications that can affect vision. The sickle cells can damage blood vessels in the retina. This is called retinopathy.

This condition can be diagnosed only by an eye specialist. Early changes may be treated. It is recommended that after the age of 5, children with sickle cell disease have their eyes examined by a specialist every year.

How Can I Help My Child Live Better?

Medical care to treat sickle cell disease has improved a lot over the years. This has increased the likelihood that children with such an illness can lead happy lives into adulthood. Parents are also better informed about how to care for their child's special needs. Here are some guidelines that can help you help your child to live a full and happy life.

9 Growth and Development

Most children with sickle cell disease grow normally when they are babies. Throughout the rest of their childhood, these children may be smaller and thinner than other children their age. Puberty may be delayed due to this slow growth. Some teenagers may be concerned about this, but they can be reassured that most often this is temporary and they will eventually catch up with their peers.

1 Know what symptoms to look for and what to do

Being well informed about sickle cell disease will increase your confidence in caring for your child. Early recognition of symptoms is the key in managing your child's illness. Your ability to seek medical care quickly when needed will help better manage some of the health problems that can occur. Not all health problems can be prevented, but may be easier to manage if detected and treated early.

Ensure that your child receives all the recommended vaccinations

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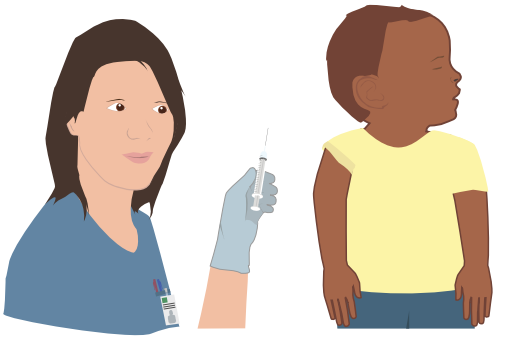


Adequate vaccination is very important for your child's health because children with sickle cell disease are more susceptible to infections. Make sure that your child gets all the recommended vaccinations as well as any other vaccines suggested by your hematology team. Bringing your child's vaccination book when you come to the clinic will help us keep track of the immunization status. Every member of the family should have a flu vaccine each year.

Encourage a healthy life style

6

A healthy life-style is important for all children, and especially for those living with sickle cell disease.

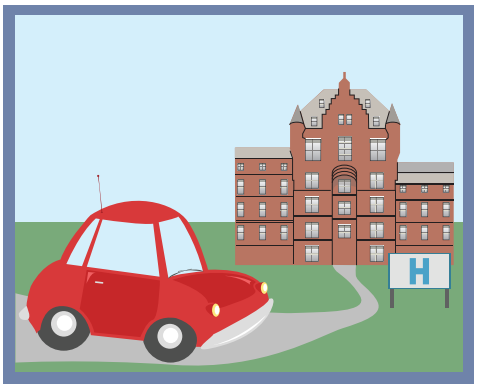
The following will help your child stay as healthy as possible:

- Eat a balanced diet
- Drink plenty of fluids
- Engage in regular play and activities
- Have time for adequate rest and sleep
- Maintain a normal body temperature by dressing appropriately for the weather

As time goes by, your child will learn to identify what are the things that make him feel good or bad. Parents need to encourage their child to engage in normal childhood activities when possible. Offering your children a healthy lifestyle will help them make better choices for themselves in the future.

Bring your child to regular medical visits

Regular medical visits are essential in the management of your child's health. Seeing your doctor or nurse on a regular basis, even when your child is well, will provide the team with valuable information that will be helpful if complications occur. It also gives you the opportunity to build a strong relationship with your hematology team, necessary in the successful management of your child's health. It will also allow you to get involved in your child's care and be part of the decision-making process. **With each clinic visit, make sure you have the proper prescriptions for the medications that your child will need until his next appointment.** If you are unable to attend a visit, we ask you to contact the secretary so she can inform the team and schedule another appointment for you.



Take care of yourself

Caring for a child with sickle cell disease can be challenging for some families. It is easy to focus only on the child and forget about one's self. It is important for you to take the time to rest as well, in order to stay healthy to best care for your child. Some parents report that a few hours of babysitting allowed them to have time for themselves. Both you and your child may benefit from this.

Some parents find it helpful to notify their employers of occasional absences. Sickle cell disease is unpredictable and when a crisis occurs, you might have to stay with your child.

If you are traveling

Traveling when you have sickle cell disease is fine. Notify your doctor or nurse of your travel plans so they can provide you with a "travel letter" and ensure that your child is properly vaccinated. This may require a visit to a specialized "Travel Clinic". Fly only in a pressurized plane and make sure to bring all the necessary medications. Offer your child plenty of fluids.

- Be careful at high altitudes
- Make sure that your child dresses according to the weather, so he doesn't feel too hot or too cold.
- Do not hesitate to consult a doctor while on your trip if your child is unwell. Don't forget to mention that he suffers from sickle cell disease.

Treatments and medications

1) Prevention

These are some of the recommendations to follow in order to keep your child as healthy as possible, and to try to prevent complications:

- **Penicillin** or an alternative antibiotic have been shown to prevent life-threatening infections in children with sickle cell disease. Your child will be prescribed penicillin, twice a day, everyday, from diagnosis until 5 or 6 years of age. Give this medication to your child every day, morning and night, even when well and do not stop until your doctor tells you so.

- **Folic acid** (folate) is a vitamin that acts as a "building block" in the production of red blood cells. Along with a balanced diet, folic acid, taken as prescribed, will help your child's body make new red blood cells.

- Ensure that your child is adequately vaccinated
- Keeping your child well hydrated will improve the shape of the red blood cells and help keep them from clogging up small blood vessels. Children with sickle cell disease need to drink more fluids than other children. Give your child fluids whenever he or she is thirsty. Your child will need to drink more when:

  - He/she has a fever
  - He/she has pain
  - He/she is very active
  - It is hot outside
  - He/she is traveling

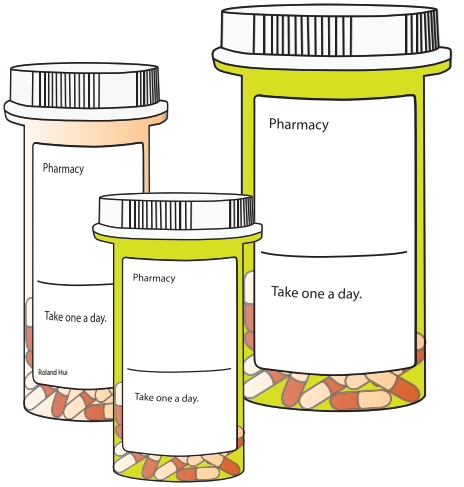
2) Treatments

- **Hydroxyurea** is another medication that may be recommended for your child. Not every child with sickle cell disease will require Hydroxyurea. To learn more about this medication, speak to your doctor or nurse.

- **Transfusions.** Under certain conditions, children with sickle cell disease may benefit from a blood transfusion. Transfusions are indicated to treat certain acute complications of the illness. They can also be used for the treatment or prevention of chronic complications and other health related events.

If your child needs to have an operation, a transfusion may be necessary beforehand to try and avoid post-operative complications. **Please notify us if your child requires surgery.**

- **Pain Medication.** There are several medications that can be used to provide prompt pain relief. Always have acetaminophen (ex. Tylenol™ or Tempra™), ibuprofen (ex. Advil™) in your medicine cabinet. Stronger medications may also be necessary to alleviate your child's pain (e.g. codeine or morphine). Keep these medications in a safe place and administer them to your child as ordered by the doctor. **Remember, pain is easier to control when treated early.**



- **Stem cell transplant** is currently the only possible cure for sickle cell disease. Not every child is a candidate for transplant. For more information on stem cell transplant, we encourage you to discuss this issue with your doctor or nurse.

Services Available to Families

Pain Management Services

Pain management can be a challenge in children with sickle cell disease. The acute and chronic pain teams may be very helpful to us in achieving the best pain management for your child. These pain management specialists will be available to you should you need them.

Genetic Counseling

Genetic counseling may be an option if you are planning to have more children. Counselors can help you to know your chances of having future children with sickle cell disease. They can also provide guidance in regards to family planning and prenatal diagnosis. We encourage you ask your doctor or nurse about genetic counseling.

Child Life Specialists

The child life specialists support the optimum continued development of infants, children and adolescents receiving hospital services by providing educational/ recreational programs that reflects normal life experiences and help to minimize psychological trauma. The child life specialists provide opportunities for play, for gaining a sense of mastery and control. They also promote learning, self-expression, peer interaction, socialization and family involvement.

Child Life Specialists may not be available in all the centers.

Nursing Assistant

The nursing assistant will see your child at each clinic visit. She will take vital signs, measure the weight and height and may assist in supervising a treatment or giving vaccinations. She works very closely with the nurse clinician and will notify her or the doctor of any changes in your child's health.

Clerical staff

The secretaries will take your calls when you need to reach a member of the team. They can help you by addressing your concerns to the appropriate health care professional. They will also coordinate your appointments to the clinic.

The Sickle Cell Association of Québec

The Sickle Cell Association of Quebec offers other services to support families, including: massage therapy, respite services, babysitting and documentation about sickle cell for children. The Association is an important source of information and promotes sharing and communication for children, parents and adults living with sickle cell disease. It's a great community resource that offers group discussions and some information on sickle cell screening as well as information on carriers. For more information, visit the website of the Association at the following address: www.anemie-falciforme.org/

Notes

Help Us Help Others

Help support the MUHC Patient Education Office! Donations make a huge difference. They help us create health information materials and programs to deliver the best care for life.

All patient materials are available, free of charge, on the internet to patients and families everywhere.

Make a donation to **MUHC Patient Education - Dr. David Fleiszer** through the Montreal General Hospital Foundation:

Online : https://www.mghfoundation.com/

By phone : 514-934-8230

By mail/in person: 1650 Cedars Avenue, E6.129, Montreal, QC, H3G 1A4, Canada

Thank you for your support!

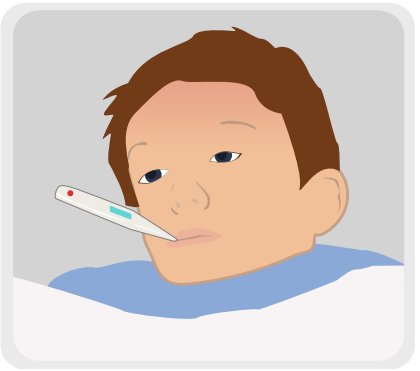
LA FONDATION DE L'HÔPITAL GÉNÉRAL DE MONTRÉAL — THE MONTREAL GENERAL HOSPITAL FOUNDATION

1821

MUHC Health Education Collection: http://infotheque.muhc.ca
MUHC Patient Education Office: www.muhcpatienteducation.ca

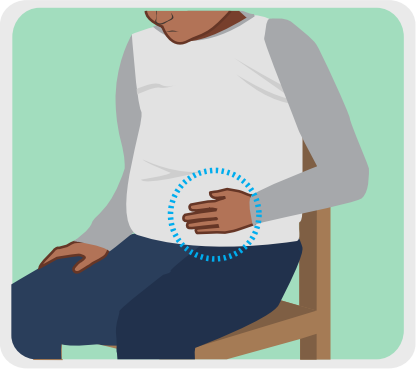
① Information Sheet: When to call and seek medical attention.

Call us and see a doctor if...



Fever

...your child has a temperature greater than 38.5°C (101°F) rectally, or 38°C (100.4°F) orally, or 37.5°C (99.3°F) axillary. A fever must not be ignored in children with sickle cell disease. **Report fever to your doctor or nurse, and take your child to the hematology clinic or to the emergency room immediately. Do not wait.**



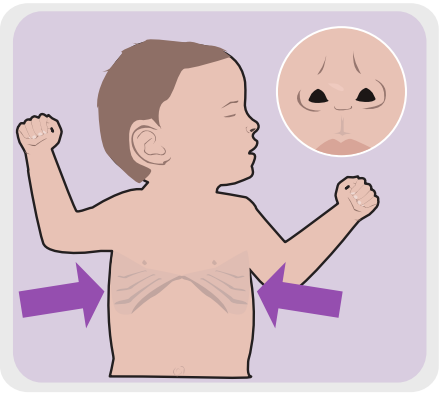
Distended or Enlarged Abdomen

...your child's belly becomes enlarged and feels hard to the touch. This might mean that the blood cells are getting trapped into the spleen, causing it to enlarge. Your doctor will teach you how to feel your child's spleen.

Prolonged pain

...your child is feeling pain that is not relieved by rest, increased fluids, Tylenol™, Tempra™, Advil™ or morphine/codeine if it is prescribed by your physician.

1 Information Sheet: When to call and seek medical attention.



Difficulty Breathing

…your child has rapid breathing, shortness of breath, persistent cough, or chest pain.

Painful and persistent penile erection

…your child (boy) develops a painful, penile erection that can last a long time (more than 1 hour).

Other symptoms

…you notice any of the following symptoms in your child:

- Headache
- Stiff neck
- Weakness or numbness of arms or legs
- Change in vision
- Severe abdominal pain

If you notice any of the above symptoms, your child needs medical care as soon as possible. Call the Hematology team and take your child to the hematology clinic or to the emergency room.

Do not forget to inform the emergency team that your child has sickle cell disease.

2 Information Sheet: Hematology Team

MCGILL UNIVERSITY HEALTH CENTRE
MONTREAL CHILDREN'S HOSPITAL

Physician:	Dr. Sharon Abish	
Nurse Clinician:	Marie Gale:	514-412-4400, ext. 22428
Nursing Assistant:	Ana Lereu:	514-412-4400, ext. 22428
Social Worker:	Laura Johnston:	514-412-4400, ext. 24455
Secretaries:	Johanne Roy:	514-412-4434 or 514-412-4400, ext. 22428
Child life specialist:	Anna Paliotti	

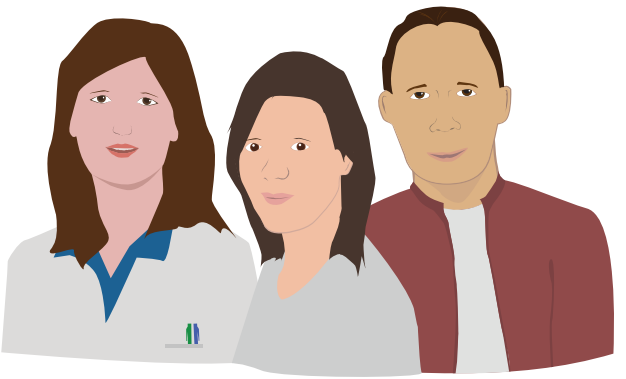
How to reach us:

Weekdays

From Monday through Friday from 8 AM to 4 PM. Call the clinic by dialing 514-412-4400, ext. 22428.

Evenings/nights or during the weekend

You can reach the pediatric Hematologist/Oncologist on call by dialing 514-412-4400, ext. 53333.



2 Information Sheet: Hematology Team

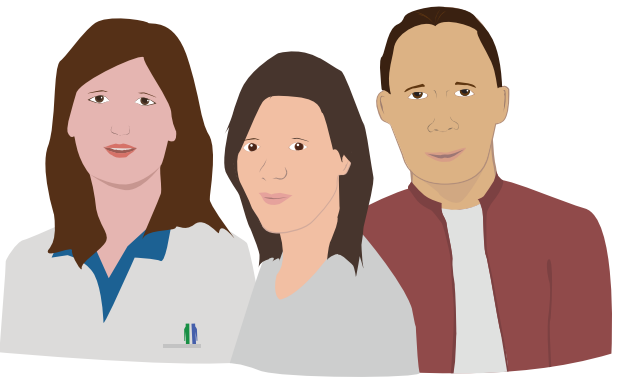
SHERBROOKE UNIVERSITY HOSPITAL CENTRE
FLEURIMONT HOSPITAL

Doctor: Dr. Josée Brossard
Pediatric hematologist-oncologist
Service of Pediatric Hematology
Department of Pediatrics

How to reach us:

Weekdays
Monday through Friday from 8 AM to 4 PM: 819-346-1110, ext. 74464.

Evenings/nights or during the weekend
Call 819-346-1110, ext. 0, ask for Dr. Josée Brossard or pediatric hematologist on call.



2 Information Sheet: Hematology Team

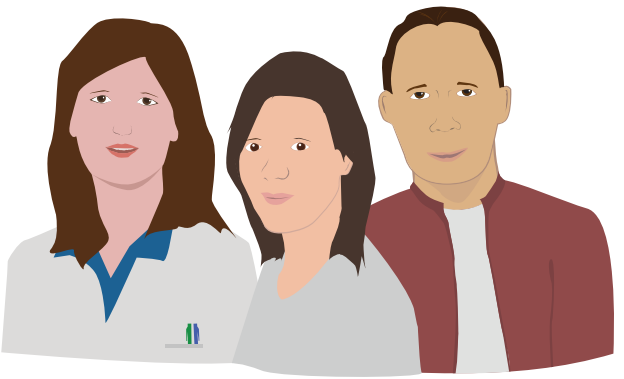
CENTRE HOSPITALIER UNIVERSITAIRE DE QUÉBEC
CENTRE HOSPITALIER DE L'UNIVERSITÉ LAVAL

Doctor: Dr. Valérie Larouche
Clinical hematology (Nursing): 418-656-4141 ext. 71418
Secretariat of Hematology: 418-656-4141 ext. 47191

How to reach us:

Weekdays
Monday through Friday from 8 AM to 4 PM: 418 656-4141, ext. 47191.

Evenings/nights or during the weekend
Call the hospital Emergency Department at: 418-654-2144.



2 Information Sheet: Hematology Team

SAINTE-JUSTINE UNIVERSITY HOSPITAL CENTRE

Doctor: Dr Nancy Robitaille
Doctor: Dr Yves Pastore
Nurse: Nathalie Fournier
Nurse: Sophie Parent

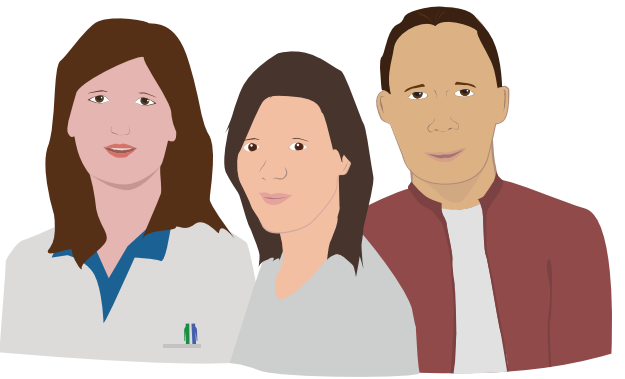
How to reach us:

Weekdays

Monday through Friday from 8 AM to 4 PM: 514-345-4931 ext. 2712.

Evenings/nights or during the weekend

After 4 PM, during the evening or over the weekend, you can call the assistant head nurse of Hematology Oncology at 514-345-4931 ext. 2111 or also by calling 514-415-6953. If you are having trouble, you can also try 514-345-4788.



3 Information Sheet: Fever

Children with sickle cell disease are more at risk of getting serious infections.

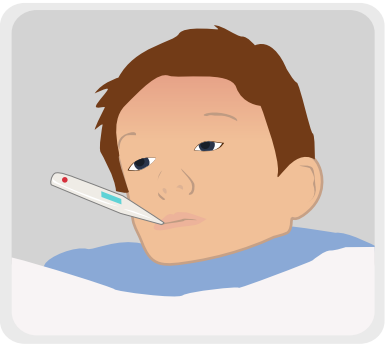
If your child has a fever, it could mean that he has an infection. You need to know how to take your child's temperature so you can report it to your nurse or doctor. If you do not have a thermometer at home, you need to get one.

If your child has a fever:

- He will feel warm when you touch him.
- His cheeks could also be flushed (red or pink in color)
- He could have chills
- He could experience headache, aching all over the body.
- He could also be irritable. You need to monitor his temperature.

Choosing a thermometer
Digital thermometers are often recommended because they are easy to read. They give accurate temperatures within 30 seconds to 2 minutes. Read the directions included, to learn how to use your thermometer.

Glass thermometers containing mercury are NOT recommended.



Temperature in the mouth (oral)
Taking the temperature in the mouth is recommended for children old enough to understand directions and be cooperative (usually 5 or 6 years old).

- Do not give your child hot or cold liquids 15 to 30 minutes before taking his temperature
- Make sure your child's mouth is clear of food, candy or gum
- Ask your child to sit down and stay still while taking his temperature
- Ask your child not to talk or bite the thermometer and to keep his lips closed together while the thermometer is in place
- Hold the thermometer in place until it beeps
- Stay with your child while taking his temperature
- Remove thermometer and read the number

3 Information Sheet: Fever

Temperature in the rectum (rectal) - For children under 2 years old

- Place your child across your lap or on something firm like a changing table
- Put a small amount of lubricant or petroleum jelly on the tip of the thermometer
- Insert the tip of the thermometer in you child's rectum about ½ an inch to 1 inch
- Hold your child firmly and keep the thermometer in place until it beeps
- Stay with your child while this is being done
- Remove the thermometer and read the number
- Clean the thermometer after each use

If you use more than one thermometer, clearly identify the rectal one to avoid placing it in your child's mouth, therefore reducing the risk of infections.

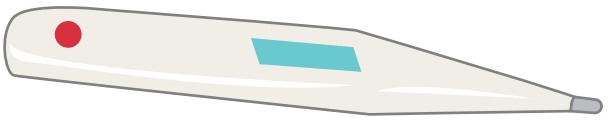
Temperature under the arm (axillary)

- Remove your child's shirt so it is easier for you to see the armpit
- Lift your child's arm and place the tip of the thermometer in the center of the armpit. Bring your child's arm down and hold it tightly against the chest to keep the thermometer in place.
- Leave the thermometer in place until it beeps
- Stay with your child while this is being done
- Remove the thermometer and read the number
- If your child is very young or uncooperative, you might want to sit him on you and hold him while taking his temperature

If your child's temperature is:

- **38.5˚C (101˚ F) rectally or more;**
- **38˚ C (100.4˚ F) orally or more;**
- **37.5˚ C (99.3˚ F) axillary or more;**

You need to see a doctor. Call the hematology team and take your child to the hematology clinic or to the emergency room. **Do not wait.**



4 Information Sheet: Pain Management

No pain (Prevention)

1. Treat any fever with acetaminophen (Tylenol™ or Tempra™). Report fever greater than 38.5˚C (101˚ F) rectally, or 38˚ C (100.4˚ F) orally, or 37.5˚ C (99.3˚ F) axillary, to your nurse or doctor.
2. Encourage your child to drink fluids regularly. This is an important way to prevent pain. Drinking fluids makes your child's blood flow better through the capillaries.
3. Dress your child well during cold weather.
4. Encourage quiet play and limit vigorous exercise for your child. Encourage breaks in between activities.

First signs of pain (Treatment)

1. Bring your child somewhere comfortable to sit or to lie down.
2. Give acetaminophen (e.g., Tylenol™) or ibuprofen (e.g., Advil) to your child as recommended on the package or as per the nurse/doctor or pharmacist.
3. Encourage your child to drink more fluid.

While waiting for the pain medication to work

1. Distract your child with a calming activity. For example, read a book or put on his favorite calming music.
2. You may give your child a warm bath, or place a warm dry compress on the painful area.
3. Use gentle massage.

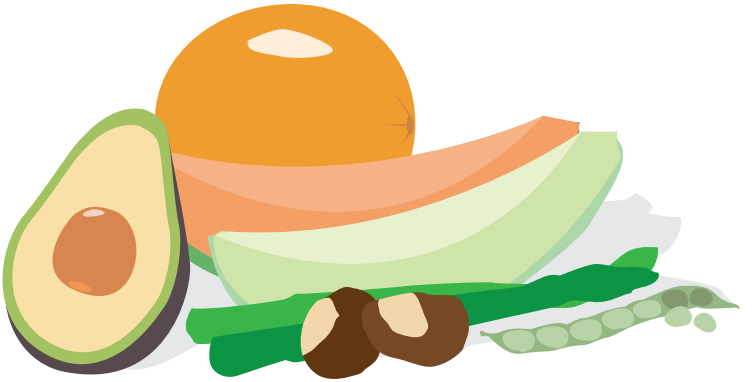
If pain does not go away within the hour

1. A stronger pain killer might be necessary. If it is not the first pain crisis and you have some codeine or morphine ordered by the doctor, give one dose to your child now.
2. If it is the first pain episode or if codeine or morphine does not decrease the pain within the next hour, call the hematology team for advice.
3. A visit to the clinic or to the emergency room for further treatment could be recommended.
4. Continue to distract your child with a calming activity while waiting to see the nurse or doctor.

5 Information Sheet: Recommended Medications

Name	How to take it	Benefits	What to look out for
Penicillin or alternative antibiotics	2 times a day, morning and night every day until 5 or 6 years of age	• Fewer serious infections	• Diarrhea • Heart burn • Nausea • Vomiting • Rash • Allergic reaction • Usually very well tolerated
Folic acid	As prescribed	• Helps your child's bone marrow make new red blood cells	• Loss of appetite
Hydroxyurea	As prescribed	• Fewer painful sickle cell crisis • Less hospitalizations • Less transfusion needs • Increases hemoglobin (regular and fetal) • Helps prevent recurrence of acute chest syndrome	• Diarrhea • Nausea • Vomiting • Skin rash • Decreased blood counts

6 Information Sheet: Food Rich in Folic Acid



Vegetables :

- Artichoke, frozen, cooked, (½ cup)
- Asparagus, cooked, 4 spears
- Beets, cooked, (½ cup)
- Broccoli, cooked, (½ cup)
- Brussels sprouts, frozen, cooked, 4 sprouts
- Edamame/baby soybeans, cooked, (½ cup)
- Escarole or endive, raw, (1 cup)
- Lettuce, Romaine or mesclun, (1 cup)
- Okra, frozen, cooked, (½ cup)
- Potato, with skin, cooked, 1 medium
- Spinach, cooked, (½ cup)
- Spinach, raw, (1 cup)
- Turnips greens or collards, cooked, (½ cup)

Fruits :

- Avocado, ½ fruit
- Orange juice, (½ cup)
- Papaya, ½ fruit

Grain Products :

- Bagel, plain, ½ bagel
- Bread, white, 1 slice
- Bread, whole wheat, 1 slice
- Cracker, saltine, 10 crackers
- Pasta, egg noddles, enriched, cooked, (½ cup)
- Pasta, spinach, enriched, cooked, (½ cup)
- Pasta, white, enriched, cooked, (½ cup)

6 Information Sheet: Food Rich in Folic Acid

Meat Alternatives :

- Beans (kidney, great northern), cooked, (¾ cup)
- Beans (mung, adzuki), cooked, (¾ cup)
- Beans (navy, black, small white), cooked, (¾ cup)
- Beans (pink, pinto), cooked, (¾ cup)
- Beans, cranberry/roman, cooked, (¾ cup)
- Lentils, cooked, (¾ cup)
- Meatless (fish sticks, meatball, chicken), cooked, (½ cup)
- Peas (chickpeas, black-eyed/ cowpeas/adzuki), cooked, (¾ cup)
- Peas, pigeon, cooked, (¾ cup)
- Soy burger/vegetarian meatloaf or patty, cooked, (½ cup)
- Soy nuts, (¼ cup)
- Soybeans, mature, cooked, (¾ cup)
- Sunflower seeds, without shell, (¼ cup)

Organ Meat :

- Giblets (chicken, turkey), cooked, (2 ½ oz)
- Liver (lamb, veal), cooked, (2 ½ oz)
- Liver (turkey, chicken) cooked, (2 ½ oz)
- Liver, (beef,pork), cooked,(2 ½ oz)
- Pate, chicken liver, (2 ½ oz)

Fish and Seafood:

- Conch, cooked, (2 ½ oz)

Miscellaneous :

- Yeast extract spread (vegemite or marmite), (2 Tbsp)

Food Sources of Folate, adapted from the Dieticians of Canada
"Canadian Nutrient File 2010".
www.hc-sc.gc.ca/fn-an/nutrition/fiche-nutri-data/index-eng.php

A balanced diet includes foods from the 4 groups as recommended in the Canada food guide. This is the best way of getting all the vitamins and minerals necessary to your child's health.

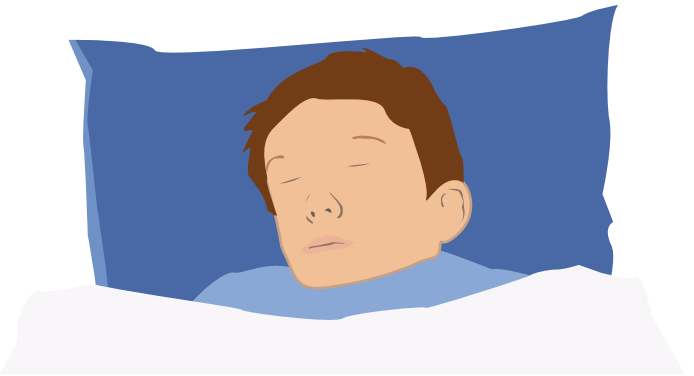
7 Information Sheet: Transcranial Doppler (TCD)

What is a Transcranial Doppler (TCD) Ultrasound ?

- TCD is a machine that uses ultrasound to detect increased blood flow in some blood vessels of the brain. Blood vessels that have been damaged by sickle cells are often more narrow and the passage of the blood through these vessels produces a louder noise. With this information, it is possible for the doctor to identify children at greatest risk of having a stroke in the future.

What is a stroke?

- A stroke is the result of a circulation problem in the brain vessels. It happens when there is a blockage, depriving part of the brain from blood and oxygen. There are many signs and symptoms associated with a stroke. Refer to the list of symptoms on page 31 of this handbook.



Why should we be concerned with strokes in patients with sickle cell disease?

- Black American adults without any hematological issues have twice the risk of white Americans to have a stroke. Approximately 11% of sickle cell patients have clinically apparent strokes before the age of 20. The risk increases by 24% by the age of 45 and is greater in the first 10 years of life, especially between 2 and 10 years old. So a child with sickle cell disease has a stroke risk that is 333 times greater than a healthy child without sickle cell or heart disease. In children with abnormal TCD ultrasounds, the risk is more than 3000 times greater.

7 Information Sheet: Transcranial Doppler (TCD)

How is a TCD ultrasounds done?

- The test is done in the radiology department of specialized centers. A secretary will call you to give you an appointment. The test a not very long (about ½ hour), although it may be a little longer in some situations especially in younger children. Your child will be awake, lying down and needs to remain still for the exam. This test is not painful and you will be able to stay with your child during the procedure.

What will happen after the test?

- The results of the TCD ultrasound will be sent to the sickle cell physician. If the results are normal, there is no need to do any treatments. This test will need to be repeated at least once a year. If the results are abnormal or questionable, we will repeat another TCD ultrasound sooner.

What will happen if the results show that my child is at greater risk of having a stroke in the future?

- If the test results are abnormal twice in a row, we will sit down with you and discuss whether regular transfusion therapy may be an option to lessen the likelihood of your child having a stroke.
- Studies have shown that regular transfusions (every 4 weeks) in children with abnormal TCD ultrasounds can help decrease the risk of having a stroke in the future.
- This test is very important. **Please make sure that your child attends the appointments!**

8 Information Sheet: General Information

Name of Child: _____

Date of Birth: _____ Place of Birth: _____

Name of Parent or Guardian: _____

Address: _____

Medicare Number: _____

In Case of Emergency

Name: _____

Relationship: _____

Main Telephone: _____ Other Telephone: _____

Doctor/Clinician

Name: _____

Telephone: _____

Birth Information

Length of Pregnancy: _____

Health during Pregnancy: _____

Caesarean: _____

Weight: _____ Height: _____

Head Circumference: _____

Problems at Birth: _____

8 Information Sheet: General Information

Development of the Child

Was your child breastfed? _____

If so, for how long: _____

Was your child given formula? _____

If so, which one? _____

When did your child start eating solid food and what was it? _____

_____

Food allergies: _____

_____

Any other problems related to food? _____

_____

When did your child start doing the following:

Speak his first words: _____

Pronounce words other than Mom and Dad: _____

Chain words together: _____

Understanding other people besides family when they talk: _____

Clap his hands: _____ Sit up without help: _____

Walk without help: _____ Run and jump on two feet: _____

Making scribbles: _____

Using index finger and thumb to pick up objects: _____

Show objects with fingers: _____

Drink from a glass: _____ Use a spoon: _____

9 Information Sheet: Growth and development

It's very important to pay attention to your child's weight and height. Write down this information with each visit to the clinic. This will help you keep track of your child's growth.

Date	Age	Weight	Height	Cranial Perimeter	Other

9 Information Sheet: Growth and development

Date	Age	Weight	Height	Cranial Perimeter	Other

10 Information Sheet: Family Medical History

Write down all the significant illnesses for each members of your close family.

Date	Relationship	Date of Birth	Health Problem

10 Information Sheet: Family Medical History

Date	Relationship	Date of Birth	Health Problem

11 Information Sheet: Allergies

Record all your child's allergies, the kind of reaction, how it was treated and at what age was the allergy discovered.

Allergy	Reaction	Treatment	Age Discovered

12 Regular vaccination schedule

Certain vaccines are recommended for everyone.
They are part of the of the regular vaccination schedule.

Vaccines protecting against:	At 2 months	At 4 months	At 6 months	At 12 months	At 18 months	Between 4 and 6	Elementary 4	Secondary 3	After 60
Diphtheria-tetanus-whooping cough-hepatitis B-polio-Hib	✓	✓	✓ (without hepatitis B)		✓				
Pneumococcus	✓	✓		✓					✓ (65 and over)
Rotavirus	✓	✓							
Flu, in autumn			✓ (6 to 23 months)						✓
Meningoccus C				✓				✓ (september 1st 2013)	
Measles-mumps-rubella-chicken pox				✓ (without chicken pox)	✓				
Diphtheria-tetaus-whooping cough-polio						✓		✓ (without polio)	
Hepatitis B							✓ (the vaccine used protects also against hepatitis A)		
Human papillomavirus							Girls only		

* A dose of Tdap vaccine is also indicated for all adults.

Other vaccinations may be recommended for reasons of health, work, activities or travel.

Santé et Services sociaux Québec

There are other vaccines recommended for children with sickle cell disease (like Menveo and Pneumovax 23). These vaccines will be administered to your child as recommended by your hematologist.

Regular vaccination schedule
Ministry of Health and Social services
http://publications.msss.gouv.qc.ca/acrobat/f/documentation/2013/13-278-16A.pdf

12 Other Vaccination and Tests

Record any other vaccinations and tests that your child has received other than the ones shown on the previous page.

Vaccination/Test	Date	Doctor/Nurse

13 Information Sheet: Hospitalizations

Record all the major injuries or illnesses that caused your child to be hospitalized, the treatment(s) he received when in the hospital, and the hospital's name.

Date	Sickness/Injury	Treatment/Medication	Name of Hospital

14 Information Sheet: Dental Health

When did your child start brushing his teeth? _____

When did your child first visit the dentist? _____

Record your child's appointments with the dentist and the treatments received.

Date	Reason	Treatment

14 Information Sheet: Dental Health

Date	Reason	Treatment

15 Information Sheet: Complete Blood Count Log

Date	WBC	Hemoglobin	Platelets	Neutro-philes	Reticu-locytes	Hb F%	Hb S%	Hb C%	Ferritin

15 Information Sheet: Complete Blood Count Log

Date	WBC	Hemoglobin	Platelets	Neutro-philes	Reticu-locytes	Hb F%	Hb S%	Hb C%	Ferritin

16 Information Sheet: Other Important Medical Appointments

Record all appointments your child had with any medical professional such as speech therapist, psychologist, ophthalmologist, social worker, physiotherapist...

Date	Reason	Name of Clinician	Profession

16 Information Sheet: Other Important Medical Appointments

Date	Reason	Name of Clinician	Profession

17 Information sheet: Guidelines to follow in case of emergencies

For patients with sickle cell disease

This protocol is for health professionals

It is meant to inform you of the treatments required, should a child with sickle cell disease present to your hospital for care

_____ suffers from sickle cell disease.

(Name)

This patient is being followed at _____.

(Name the center)

Summery :

Sickle cell disease is a serious illness that may lead to severe complications. Here are some guidelines to follow in order to ensure proper management of these potential complications.

1. Fever, chest pain and respiratory distress, are serious symptoms in a child with sickle cell disease.
2. All patients known to have sickle cell disease, who present with any of these symptoms, must be triaged quickly and seen by a physician immediately upon arrival to the emergency room.
3. In addition to the usual vital signs, monitoring of oxygen saturation in room air is essential.
4. If the child is febrile, start antibiotics intravenously immediately after blood cultures have been drawn. Do not wait for the chest x-ray.
5. Pediatrics consultation for all patients.
6. Notify the hematology team of the center where the child is being followed as soon as possible.
    - The information sheet called “The hematology team” (p.47-8-9 and 50 of this guide) contains the numbers of all four designated centers.

17 Information sheet: Guidelines to follow in case of emergencies

For patients with sickle cell disease

Fever ≥38.5◦C rectal or ≥38◦C oral or 37.5◦C axillary

NOTIFY THE DESIGNATED CENTER RIGHT AWAY

Nursing care

- Monitor vital signs, including temperature and oxygen saturation, every 4 hours
- Notify physician if O_2Sat ≤94% , if the patient experiences difficulty breathing or unusual sleepiness or if RR<10/minute **or** >40/min
- Administer O_2 to keep saturation >94% (unless the patient's baseline saturation is normally lower)
- Notify physician if patient is unstable

Investigation

- CBC with reticulocytes count followed by daily CBC
- Electrolytes, urea, creatinine, LDH, AST, ALT and total/direct bilirubin every second day
- Blood cultures
- Chest x-ray (to be done after the first dose of antibiotics has been given)
- Urine analysis and culture (to be done automatically if the child is younger than 3 years old and only if symptomatic in older children)
- Throat culture PRN (depending on clinical evaluation)

Treatments

- D5% NaCl 0,9% at _____ mL/h (1X maintenance)
- Incentive spirometry : 10 deep inspirations / every 2 hours while awake
- Ibuprofen _____mg (10 mg/kg/dose, max 400mg) PO QID regularly
- Acetaminophen : _____mg (15mg/kg/dose, max 650mg) PO every 4 hours PRN

IV antibiotics

- To be started as soon as possible
- Ceftriaxone _____mg (50-80mg/kg/dose, max 2000mg) IV or IM every 24 h. If given IM, reconstitute vial with lidocaïne 1%.
- If chest x-ray shows infiltrates:
  - o add Clarithromycin_____mg (7,5 mg/kg/dose, max 500mg) PO every 12 hours
- If patient is allergic to cephalosporins:
  - o Vancomycin _____mg (10mg/kg/dose, max 500 mg) IV every 6 hours

17 Information sheet: Guidelines to follow in case of emergencies

For patients with sickle cell disease

Pain Crisis

NOTIFY THE DESIGNATED CENTER RIGHT AWAY

**Protocol to be initiated only after the diagnosis of pain crisis has been established and other health problems have been ruled out.*

Start with:

- Ibuprofen _____mg (10 mg/kg/dose, max 400mg) PO QID regularly
- Acetaminophen : _____mg (15mg/kg/dose, max 650mg) PO every 4 hours PRN
- IV hydration : NaCl 0.9% at _____ ml/h (1,0 x maintenance)
- Heat

Pain score (scale 0 to 10)

- Moderate pain <5/10
  - o Morphine _____mg (0,3 mg/kg/dose, max 15mg) PO every 3 hours regularly
- Severe pain >5/10
  - o Morphine _____mg (0,1 mg/kg/dose, max 10mg) IV over 20 minutes every 3 hours regularly
- Breakthrough dose for unrelieved pain :
  - o Morphine _____mg (0,05 mg/kg/dose, max 5mg) IV every hour PRN
- If more than 2 breakthrough doses are necessary over a period of 6 hours, increase dose of morphine given regularly :
  - o Morphine _____mg (0,15 mg/kg/dose, max 10mg) IV over 20 minutes every 3 hours regularly
- Hypersensitivity to morphine :
  - o Hydromorphone _____mg (0,015 mg/kg/dose, max 2mg) IV every 4 hours regularly

18 List of Resources and Websites

www.cdc.gov/ncbddd/sicklecell

http://scinfo.org/

www.stjude.org/sicklecell (downloadable lit.)

www.anemie-falciforme.org

www.nih.gov (management of sickle cell disease)

“Teenagers living with sickle cell disease” (available in clinic)

“What is sickle cell anemia” (comic strip - available in clinic)

“Hope and destiny: The patient and parent’s guide to sickle cell disease and sickle cell trait” by Allan F. Platt Jr.

Do not hesitate to ask a member of the treating team for any specific information you might need.