

Why is Phenotypical blood better for me?

Phenotypically matched blood means that the blood has been matched to your blood group as closely as possible. This reduces the chance of having a transfusion reaction and of developing an Antibody to the blood. However, because most blood donors are from ethnic backgrounds different to Sickle Cell Disease patients, there is a limited supply of suitable blood.

How can I help increase Phenotypical blood reserve?

The Canadian Blood Service (CBS) would like more blood donors from the ethnic minorities. If you have friends or family who want to help people with Sickle Cell Disease, becoming a blood donor, is a very practical, valuable and worthwhile way to help. If you would like to arrange a blood donor drive or session, the CBS can help with this. You cannot donate blood if you have Sickle Cell Disease, and if you have Sickle Cell Trait you can donate, but the blood would not be used for a patient with Sickle Cell Disease.

Is it important to have a specialist OR is my family doctor sufficient to take care of me and why is it important to ask questions?

It is very important that you have a Family Physician who can provide day to day and long term care to you, and to look after any other health related issues. It is also important that you are seen by a specialist at least once a year to make sure there are no complications or serious problems with the Sickle Cell Disease and to be able to offer advice on treatments, and other complex issues, such as family planning and genetic testing. It is also very helpful to belong to and participate in patient advocacy groups as these are a great source of advice and assistance to individuals, as well as their role in advocating for patients as a whole.

It is important to understand your health and condition, as this then lets you take charge of your health and cope with the Sickle Cell Disease better. It can often be useful to write down questions before seeing the doctor, as many people forget what they want to ask, once in the consultation room.

What is the cure to Sickle Cell Anemia?

At present, there is only 1 cure for Sickle Cell Disease – Bone Marrow Transplant. This is limited mainly to children with severe Sickle Cell Disease. There is a lot of research into gene therapy and new drug treatments, but these are experimental at the moment.

Why are Clinics and Hospital Admissions important?

Although it is often felt that the care received in ED is not great, if you are having a bad crisis, it is safest for you to go to hospital and get checked out. There are dangers of taking too many painkillers (accidental overdose) and also the crisis could be more serious than you think. It is also important to keep in

touch with your healthcare team when you are well: so that they can help you stay healthy and deal with any complications.

Are you a member of Sickle Cell Awareness Group of Ontario?

Sickle Cell Awareness Group of Ontario (SCAGO) is a membership based not-for-profit, charitable organization with the mission of educating the community in order to have the incidence of sickle cell births reduced.

It also provides support in various ways to the patients to help ensure their utmost mental, emotional and physical.

We at SCAGO understand the toll that Sickle Cell Disease has on the patients and their family members. This is why it has created various supportive programs such as Monthly Learning for life Seminars; the SCAGO Youth Group (for ages 18-25) and the Parents Support Network. Being a part of a supportive and educational group is important to the overall mental and physical health of patients and their caregivers. We encourage you to join one of SCAGO'S supportive groups.

Charitable Registration #: 83332 0872 RR0001



SCAGO is a member of:

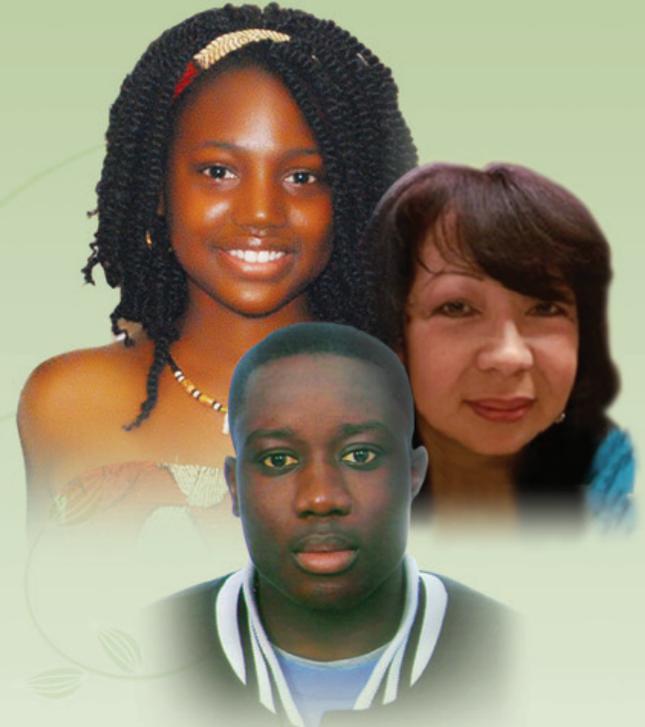
- Canadian Organization for Rare Disorders (CANADA)
- National Organization for Rare Disorders (USA)
- Black Health Alliance (BHA)

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So I have Sickle Cell Anemia



For individuals living with Sickle Cell Disease, knowing as much as possible about the disease will help to better manage and prevent/reduce complications. Sickle Cell Awareness Group of Ontario works closely with health care professionals & the Ministry of Health to ensure that individuals living with Sickle cell disease receive the best care possible.

Stay well. Stay hydrated. Stay positive. Stay connected to a group.

What is Sickle Cell Anemia?

Sickle Cell Disease (also called Sickle Cell Anemia) is a condition you have inherited and are born with. It is caused by an abnormal form of Hemoglobin. This is the part of your Red Blood Cell that carries oxygen around the body and keeps your vital organs working. With Sickle Cell Disease, the abnormal Hemoglobin (HbS) is not able to work properly. The Red Blood Cells become stiff and block up the blood vessels in your body, causing pain and damage, and they also get destroyed quickly, leading to anemia.

What does Sickle Cell Anemia mean to me?

It is something you will have your whole life, and need to learn how to live with it, so that you can continue living as normally as possible, with schooling, work, and family life. As it is inherited, you also need to know your partner's Sickle Cell status as you could pass it onto your future children too.

What are the complications to look for?

Sickle Cell Disease causes problems in 2 ways, by the breakdown of the red blood cell (hemolysis) and by blocking the flow of blood in the blood vessels (vaso-occlusion). This most commonly causes pain, especially in the bones. Other painful complications include priapism (see below), damage to shoulder and hip joints (avascular necrosis), chest pain (acute chest syndrome). There are also a lot of problems that may not be painful. These include damage to the lungs, heart (heart failure, pulmonary hypertension), kidney, liver and eyes, and also stroke, leg ulcers, infections. Because it can damage all of these body "systems", Sickle Cell disease is an example of a "multisystem disorder".

How can I prevent developing these complications?

Many complications can be prevented or reduced by either regular blood transfusions or Hydroxyurea tablets. However, both of these have side effects and there are simple lifestyle changes that can make a big difference. These include regular exercise, a good diet, not smoking and not drinking too much. Reducing the amount of stress in your life is also important, as is avoiding things that can provoke a painful crisis. Making sure you are up to date with vaccinations can help to prevent serious infections.

By seeing your doctor in clinic, when not in crisis, you have an opportunity to discuss these ways of coping with Sickle Cell Disease, and reducing complications.

How do I manage the frequent pain (crisis) that affects my chest, legs, and arms from time to time?

The best way to treat a pain crisis, is to try and avoid it starting in the first place. Try to keep warm in cold weather, and not getting dehydrated in the heat of summer. Avoid and reduce any stresses in your life as much as possible and look after your health. If you feel a crisis coming on, try to rest and drink

plenty of fluids and use your pain killers/analgesia. It is easier to stop a pain crisis getting worse if you hit the pain hard when it first comes on. If you are getting pain in the same part of the body each time, there may be something else going on, and you should see a doctor. If you are getting a lot of pain episodes (having to go to ED >3 times a year) then it may be worth trying Hydroxyurea.

What is Priapism?

This is a painful crisis affecting the penis, so only occurs in boys and men. The penis can be very hard and painful and you are unable to stop the erection. It may last a short time and happen frequently (stuttering), or you may get just one attack which is lasting for several hours (fulminant). It often comes on at nighttime when the bladder is full.

What are the treatment options for Priapism?

You can try to prevent priapism by urinating before going to bed at night, and if you wake during the night. Treat an attack like any other painful crisis: fluids, pain relief and keep warm. You may find a warm bath or shower can help. Some people can stop the attack by self-ejaculating/masturbating. If you get several episodes, Pseudofed tablets can help to prevent further attacks. If the priapism is lasting more than 30 minutes, you should go to ED as there is a risk of permanent damage to the penis and impotence. In ED they may try the measures suggested above, or may try to remove the sickled blood from the penis with a needle and syringe. This can very quickly end the priapism and feel better. If you have a lot of problems with priapism, you should see an Urologist for further advice, and also consider Hydroxyurea treatment.

I have been told to remove my spleen. Why?

We often do not recommend removing the spleen, and most adults with Sickle Cell Disease have a shrunk spleen which causes no problems. In children, and sometimes in adults, there can be a crisis where all your blood suddenly gets trapped in the spleen (sequestration crisis). This can be life threatening, and if it happens more than once, it may be recommended to remove the spleen to stop it happening again. The surgery (Splenectomy) can often be done by keyhole surgery (laparoscopic) and is a fairly straightforward operation. As the spleen helps fight infection, you will be immunized before the surgery.

How do I prevent/reduce the incidence of Crisis and other complications/symptoms associated with Sickle Cell Disease?

Both regular blood transfusions and Hydroxyurea therapy can reduce pain and other complications of Sickle Cell Disease. However, the best way is to try to prevent problems, by looking after your body and taking care of your health.

What is Hydroxyurea?

Hydroxyurea is a drug (capsule) that was originally used to treat

some forms of blood cancer (Leukemia) and blood disorders that can later turn into Leukemia. It has been used for over 15 years to treat Sickle Cell Disease.

Hydroxyurea works in a few different ways, but the main way is to increase your body's production of Fetal (baby) Hemoglobin. We know that this then protects you against Sickle Cell Disease. It is a safe drug and there is no evidence to suggest increases the risk of cancer in patients with Sickle Cell Disease. Like all drugs, it does have some side effects, and you need regular blood test monitoring. Also, unlike pain killers which you take when required, Hydroxyurea must be taken every day to be effective.

Hydroxyurea is the only drug therapy available for Sickle Cell Disease and has been shown to prolong survival, reduce pain episodes and reduce some of the complications. Unless you have been told by a specialist that you have very mild Sickle Cell Disease, you should consider going on the drug.

Why am I being offered this treatment?

You are most likely to be offered Hydroxyurea because you have lots of painful crisis or Acute Chest Syndrome. It may also be offered if there is concern about damage to any of your body organs from the Sickle Cell Disease, or as an alternative to blood transfusion treatment.

What is the difference between Blood Transfusion and Blood Exchange therapy?

Blood transfusions in Sickle Cell Disease can be carried out in 1 of 3 ways. The easiest way is to simply transfuse units of blood, or a "top-up transfusion". This improves the supply of oxygen to your body. The most complex way of receiving blood is by "automated red cell exchange" (erythrocytapheresis). This involves hooking you up to a machine that automatically filters of the Sickled Red Blood Cells and replaces it with new, normal Red Blood Cells. The alternative to these, is a "partial manual exchange transfusion", whereby a nurse will remove 2 units of blood and then give you a 2 unit transfusion. These 3 methods all have advantages and disadvantages and are suitable for different reasons.

When do I need Blood Transfusion and/or Blood Exchange therapy?

Transfusions are most commonly given for patients who have had a stroke or are found to be at high risk of having a stroke. It can also be given for a severe crisis, such as Acute Chest Syndrome or if you are very sick and need to be cared for in Intensive Care. You may need a blood transfusion if you are very anemic (aplastic crisis), which is usually due to a viral infection (cough or cold). It may also be appropriate to give you a transfusion if you are pregnant and there are complications, or if you are planning on having major surgery.