



**So my
child has
Sickle Cell
Disease...**

SCAGO

Sickle Cell Awareness Group of Ontario



Sickle Cell Awareness Group of Ontario (SCAGO)

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So... my child has Sickle Cell Disease

Your child has been diagnosed with Sickle Cell Disease (SCD), either by Newborn screening or by investigations for a medical problem. Knowing as much as possible about the disease will help you to better manage and reduce complications.

The Sickle Cell Awareness Group of Ontario (SCAGO) works closely with health care professionals and the Ontario Ministry of Health and Long Term Care to ensure that individuals living with SCD receive the best care possible.

Normal red blood cells Front and side view



Normal red blood cells are disc shaped and squishy. They flow easily through even small blood vessels with little difficulty.

Sickle red blood cells



Sickled red blood cells are crescent shaped and stiff. They break easily and can stick to the walls of blood vessels and to other cells. If blood vessels become blocked, the tissues they feed cannot get oxygen and nutrients.

What is Sickle Cell Disease?

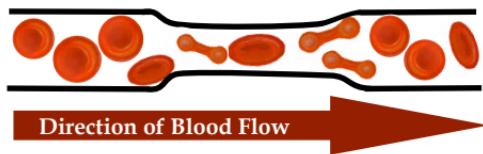
Sickle Cell Disease is a genetic disease that affects hemoglobin in red blood cells. Hemoglobin is a protein that carries oxygen to all organs in the body.

SCD is a genetic disease that is passed on to a child from both the mother and the father of the child. Most parents who have a child with SCD don't know they carry the gene for the disease because they have the trait - they have a normal hemoglobin gene, and a gene associated with sickle cell disease. Parents who carry a single gene associated with SCD have no symptoms. If a child gets a SCD gene from each parent, they will have the disease. Sickle cell disease is not infectious; you cannot catch it from someone.

SCD causes red blood cells to become stiff and crescent-shaped. These sickle cells are fragile; they break easily as they flow through blood vessels. They do not survive as long as normal red blood cells. People with SCD are anemic. This means they may look pale and feel weak. Sickled red cells stick to each other and to blood vessel walls. This can block the flow of blood to different parts of the body.

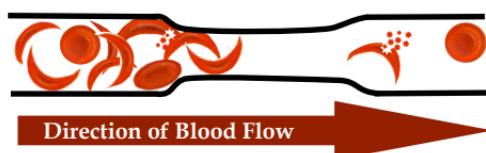
When blood vessels become blocked, pain and tissue injury can occur. Some tissues may become permanently damaged.

Normal Blood flow



Normal red blood cells are soft and squishy, like water-filled balloons. They can flow freely through small blood vessels, even if there is a small narrowing, with little risk of breaking.

Sickle Blood flow



Sickled red blood cells are hard and stiff. They stick to blood vessel walls and block the flow of blood. Sickled red cells can also break, decreasing their numbers and causing anemia.

Understanding the genes

There are different types of sickle cell disease. This depends on which genes have been passed from the parents to the child. The most common scenario is when both parents have sickle cell trait (AS). With each pregnancy, each parent will either pass the A or the S gene to his or her child.

The most common types of SCD are:

> **Sickle cell anemia or SS disease** where a child inherit an S gene from each parent.

> **Sickle-hemoglobin C or SC disease** where a child inherits an S gene from one parent and a C gene from the other

> **Sickle-thalassemia disease** where a child inherits a S gene from one parent and a thalassemia gene from the other.

Parents with a trait usually have no symptoms. You will only know you carry a gene associated with Sickle Cell Disease if you do a simple blood test. Your family doctor can help you with this.



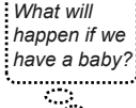
This child has Sickle Cell Disease. He got an S gene from each parent

SS

When both parents have the trait, each parent will pass on either a S or an A gene.



This child has Sickle Cell Trait. He got an S gene from mom and an A gene from dad.



This child has Sickle Cell Trait. She got an S gene from dad and an A gene from mom.

AS



With EACH pregnancy, there is a 25% chance the baby will have Sickle cell disease.

AS AS



This child has Normal Blood. She got an A gene from each parent

AA

What are some of the problems in sickle cell disease?

- > **Anemia** - children with SCD have a low red blood count. This makes them tired easily.
- > **Jaundice** - a yellow colour usually seen in the white of the eyes. This is usually a mild and common problem.
- > **Infections** can occur more often in children with SCD. This is because their spleen (an organ in the tummy that filters blood to remove germs) does not work as well. If your child has a fever, it could be a sign of a serious problem. You should contact your doctor for advice.
- > **Acute splenic sequestration** - this happens when red blood cells become trapped in the spleen. The spleen can become very large. If too much blood becomes trapped, an urgent blood transfusion may be needed.
- > **Pain** is most often felt in the bones and joints but can happen in any part of the body. It happens when sickled red cells block blood flow to the tissues. This prevents oxygen and food getting to the tissues. Young children may get dactylitis which is painful swelling of the hands and feet.
- > **Acute chest syndrome** - this is similar to



pneumonia (infection in the lungs). It can be treated in hospital.

> **Peeing often** - Children with SCD drink a lot of water as it makes them feel better. They also make more urine (pee) so they need to go to the toilet often.

> **Stroke** - Symptoms of a stroke include, weakness in the limbs, slurring of speech or being confused. These can be signs that blood flow to the brain is affected. This is a serious problem that needs to be treated quickly.

Many of these problems are preventable so it is very important to keep your follow-up appointments with your family doctor and hematologist.

Can Sickle Cell Disease be cured?

At present, there is only one cure for SCD – bone marrow or stem cell transplantation. This is a potentially dangerous procedure that requires identifying a well-matched donor, and is usually only performed in children. There is a great deal of progress being made in gene therapy, however, and if successful this will allow for cure in a much larger number of patients than are currently offered bone marrow or stem cell transplantation.

How is sickle cell disease managed?

This first thing you can do is to make healthy lifestyle changes for you and your child:

> Eat a balanced diet with lots of fruit and vegetables. Your doctor may also prescribe folic acid (a vitamin), which helps to make new red blood cells.



- > Drink lots of water - good hydration is essential.
- > Keep warm.
- > Keep your doctor's appointments.

Other important things to remember:

Prevent infection:

Most children with sickle cell disease will need to take daily antibiotics. This is to help prevent serious infections. Some children can get fever even when taking antibiotics. If this happens speak to your doctor. Your child will also need special immunizations. Your hematologist will advise you of this.

Screening:

Screening helps find problems before they become serious. These may include regular blood tests to check on your child's blood count. There is also a test called Transcranial Doppler or TCD. TCD helps to find children at highest risk for a stroke.

Medication:

Your doctor may prescribe a drug called hydroxyurea (hi-drox-ee-U-ree-ah). Hydroxyurea increases fetal (baby) hemoglobin also known as Hemoglobin F (HbF) in red cells. This means they are less likely to sickle and block blood vessels. Children on hydroxyurea will need to be watched closely. It is important to keep doctor's appointments.

Surgery

Some children will need to have their spleen removed if they have more than one episode of acute splenic sequestration. Children can grow and thrive even without a spleen but they will need to take antibiotics for a long time.



Other treatments:

Some children will need regular blood transfusions to prevent complications of the disease. Oxygen, IV fluids and referral to other health specialists such as a physiotherapist, a dietician or psychologist may be helpful.

Remember...

Every child is different. Go to prescribed clinics and work closely with your child's specialist. Both you and your doctor will be able to understand how this disease will affect your child. You will also be able to find interventions that will help your child best.

What else can you do?

Children with sickle cell disease may miss school because of their health. Talk to teachers

to make sure your child doesn't fall behind. Good education is needed for any child with a chronic illness (a disease that last for a long time) such as sickle cell disease. Most people living with SCD lead healthy and productive lives. Many go on to complete school and have families of their own. With your help, your child can achieve their full potential!

Good to Know!

Important Facts about Blood transfusions

Why is phenotype-matched blood better for me?

Phenotype-matched blood means that the blood has been matched to you much more carefully than is regularly performed. This reduces the chance of having a transfusion reaction and of developing an antibody to the blood you are transfused. However, in Canada, because most blood donors are from ethnic backgrounds different than SCD patients, there is a limited supply of phenotypically-matched blood.

How can I help increase phenotype-matched blood reserve?

Canadian Blood Service (CBS) is looking for more blood donors from ethnic minorities. If you have friends or family who want to help people with SCD, 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 in Ontario, SCAGO can help with this. Note that you cannot donate blood if you have SCD. However, individuals with sickle cell trait can still donate.



What is SCAGO?

The Sickle Cell Awareness Group of Ontario (SCAGO) is Ontario's largest patient support and advocacy organization providing psycho-social and life improving services to families with children, adolescents, and adults, with sickle cell disease. Since 2005, SCAGO has been creating and delivering evidence-based supports and services across Ontario with the goal of enhancing the quality of life of those affected by SCD, and reducing the number of new births in the province. It also advocates on their behalf with the government, schools and the health system for rightful accommodations including excellent standard of care by the medical professionals.

To learn more about our services, visit our website at: www.sicklecellanemia.ca or
E-mail us at info@sicklecellanemia.ca

Membership

SCAGO is a member of:

Sickle Cell Disease Association of Canada/ Association d'anémie falciforme du Canada (SCDAC/ AAFC)

Canadian Organization for Rare Disorders (CORD)

National Organization for Rare Disorders (NORD- America)

The contents of this booklet are for general medical information purposes only.
Always check with your doctor if you have specific questions or for further clarification of the contents of this booklet.