Alberta Health Services
Alberta Newborn Screening

Talking with Parents > When talking to a parent whose baby has received an abnormal screen result, let them know that an abnormal screen result doesn't mean that the infant has the condition. It means that they have a higher chance of having the condition, and they need more tests to find out for sure. A printable sheet for parents with more condition specific information can be used to support your conversation, visit ahs.ca/nms and under Quick Reference click on *What conditions are screened for?*

Sickle Cell Disease (SCD)

Information for Health Professionals

Also known as:

- sickling disorders
- HbS disease
- sickle cell anemia
- S, beta-thalassemia
- hemoglobin S,C disease

What is SCD?

Sickle cell disease (SCD) is an inherited disorder that affects hemoglobin. Under certain conditions, the red blood cells (RBC) acquire a crescent or 'sickle' shape. Sickle cells are prone to premature breakdown and get trapped in blood vessels causing pain, tissue damage and anemia.

What causes SCD?

The main form of hemoglobin is made up of two beta-globin and two alpha-globin chains. SCD is caused by abnormalities in beta-globin. There are a number of different beta-globin gene variant combinations that can cause SCD. The most common disease causing variant combinations are two sickle cell variants (Hemoglobin SS), a sickle cell variant paired with a beta-thalassemia variant (Hemoglobin S β-thalassemia), and a sickle cell variant paired with a hemoglobin C variant (Hemoglobin SC).

How common is SCD?

SCD occurs in all ethnic groups and is more common in certain populations such as African, Mediterranean, Middle Eastern and Asian. In some communities the incidence of SCD is as high as 1 in 400.

What are the clinical features of SCD?

Infants with SCD appear normal at birth and may have normal bloodwork. They are more susceptible to infection, sepsis, life threatening anemia and respiratory distress. Infants with SCD can also develop painful sickle crises, particularly in their hands and feet, and are prone to sickling in their spleen or splenic sequestration.

What is the screening test for SCD?

Screening for SCD is performed by examining hemoglobin patterns by chromatography. Newborn screening will not detect all infants with SCD or other hemoglobinopathies. Infants with symptoms of SCD need timely assessment and diagnostic testing even if their screen result is normal.

How is the diagnosis of SCD confirmed?

The diagnosis of SCD is confirmed by molecular genetic analysis. Hemoglobinopathy investigations may be performed. Hematology Clinic will arrange diagnostic testing.

How is SCD managed?

SCD is managed with prophylactic antibiotics, immunizations, and emergent care for fevers. Management of anemia and pain are important components of treatment. Infants will also have ongoing surveillance and management of conditions associated with SCD. Early treatment with prophylactic antibiotics reduces the risk of serious infection and overwhelming sepsis. Early educational intervention regarding triggers for acute anemia, pain episodes and splenic sequestration improves prognosis.

Is SCD inherited?

SCD is inherited as an autosomal recessive disorder. Parents of a child with SCD are carriers of the condition and have a 1 in 4 chance of having another affected child in each subsequent pregnancy. Genetic counselling is available to families with SCD.

Additional resources are available through:

Hematology Clinic (Edmonton)

Stollery Children's Hospital
8440 – 112 Street
Edmonton, AB T6G 2B7
Tel: 780-407-2654
Fax: 1-888-775-9068

Emergency consultations:
Phone 780-407-8822 and ask for the pediatric hematologist on call.

Hematology Clinic (Calgary)

Alberta Children's Hospital
28 Oki Drive NW
Calgary, AB T3B 6A8
Tel: 403-955-7155
Fax: 403-955-7393

Emergency Consultations:
Phone 403-955-7211 and ask for the hematologist on call.

Early screening and follow-up care – every baby, every time

For more information about the Alberta Newborn Screening Program, visit www.ahs.ca/newbornscreening