

Sickle Cell Disease (SCD)

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. Specialists at the clinics listed below will arrange diagnostic testing.

How is SCD treated?

SCD is treated with prophylactic antibiotics, immunizations, and emergent care for fevers. Management of anemia and pain are important components of treatment.

What is the outcome of treatment for SCD?

Early treatment with 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 and prenatal testing are available to all families with SCD.

For additional resources, please call:

Hematology Clinic (Edmonton)

Stollery Children's Hospital
8440 – 112 Street
Edmonton, AB T6G 2B7
Tel: 780-248-5415
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, detection and treatment – every infant, every time

For more information about the NMS Program, visit www.ahs.ca/newbornscreening
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