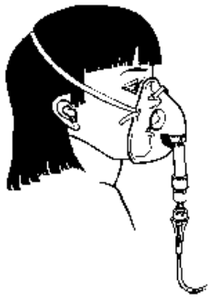
Since one of the common causes of a sickle-cell crisis is the lack of oxygen in the body, giving oxygen to children offers some degree of pain relief and it prevents the crisis from worsening.



Fluids

Being even slightly dehydrated gives the body enough stress to cause a painful crisis.

To prevent this from happening, people are given more fluids through an intravenous line. These fluids also have the right amount of minerals and sugar which the body needs.

Drinking more liquids like water and juice greatly helps too.

Blood transfusion

Blood transfusions are sometimes ordered by the pediatrician when the amount of blood in the body drops too low.

Hydroxyurea

Hydroxyurea is the first approved drug for the management of sickle-cell anemia. It was shown to decrease the number and severity of attacks in clinical studies.

This drugs is however, relatively new and is not suitable nor works for each sickle-cell case.

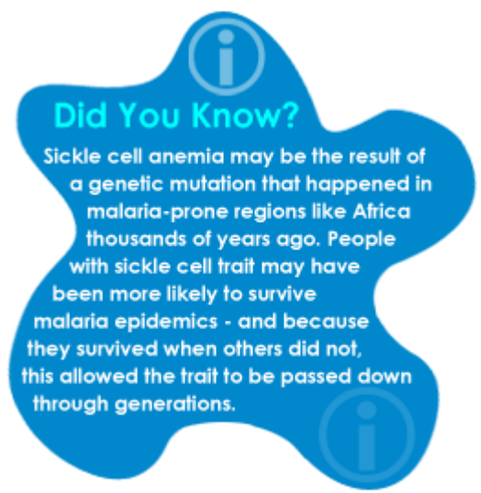
Prevention is the key!

With good health care, many people who have sickle cell anemia can live productive lives. They also can have reasonably good health most of the time and live longer today than in the past.

You can reduce the risk of a crisis occurring by:

- Drinking enough fluids
- Getting enough fresh air
- Quickly treating infections
- Avoiding extremes of heat and cold
- Dressing warmly at all times
- Avoiding emotional stress
- Avoiding smoking
- Getting proper immunization to prevent infection-triggered crises

Take medication as prescribed by your doctor.



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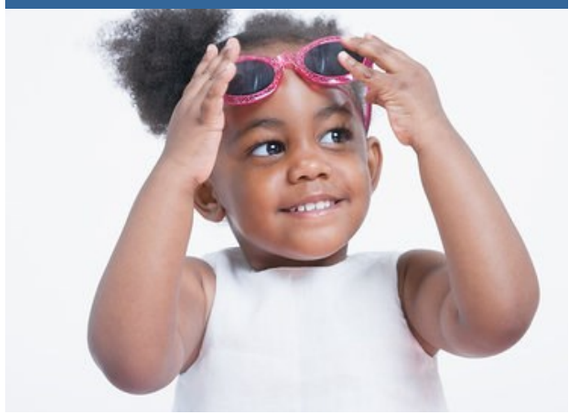
Sickle-cell Anemia



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What is sickle-Cell Anemia?

Sickle-cell Anemia is a life-long disease of the blood that makes the normally round red blood cells turn stiff and sickle-like in shape.

1 in every 1,000 live births may develop sickle-cell anemia in St. Maarten. Signs of sickle-cell anemia is usually present themselves in childhood.

It is commonly seen in people from South and Central America, the Caribbean, the Middle East and those of African-descent.

What happens in Sickle-Cell Anemia

Hemoglobin is the part of a Red Blood Cell that carriers oxygen. **Hemoglobin A (HbA)** is found normally in most people and does not cause any harm. But a certain type of hemoglobin causes Sickle-cell Anemia and it is called **Hemoglobin-S (HbS).**

When the person is under a low-oxygen environment, or when it gets too cold, the HbS changes the normally-round blood cells into something that look like crescents or **sickles.** These fragile, sickle-shaped cells deliver less oxygen to the body. They also clog the small veins in the body and stop a healthy blood flow. This causes pain. A laboratory test is needed to determine if a person has this HbS in their body.

Normal Red Blood Cell

Sickle Cell

How do I get it?

When both parents have HbA; their children would not have the sickle-cell disorder.

A child may have Sickle-cell Anemia only when **both parents** have an abnormality in their blood.

If one of the parents has the HbS, there is a chance that one of their children would carry the Sickle-cell trait, but would not get sick with it. This is what we call a **Carrier.**

But when both parents are Carriers, there is a high chance that their children would get sick with Sickle-cell Anemia.

Carriers and those positive with Sickle-cell Anemia may pass this disease to their children, starting the chain again.

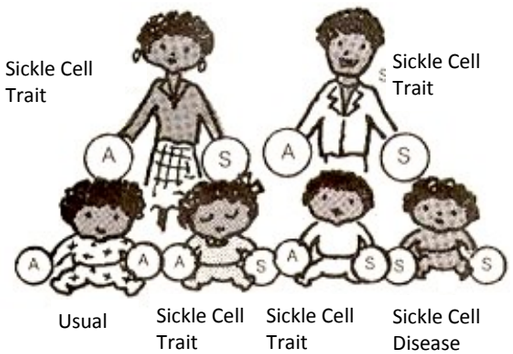


Diagram showing how the normal hemoglobin-A (HbA) and the abnormal hemoglobin-S (HbS) is passed by parents. Both parents are carriers.

A Crisis!

Almost all patients with sickle cell anemia have painful episodes called **crises**, which can last from hours to days. These crises can affect the back, arms, legs and/or the chest.

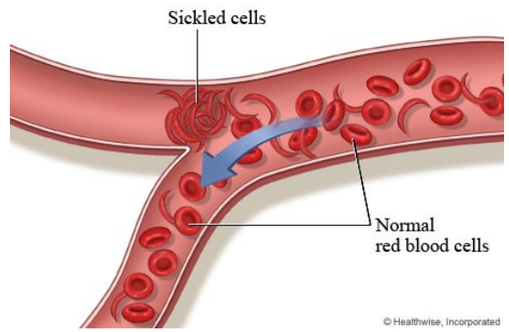
Some patients have one episode every few years. Others have many episodes per year. The crises can be severe enough to require a hospital stay.

Here are two common types of Crisis:

The Vas-Occlusive Crisis:

This occurs when an organ's blood supply is blocked by sickled-cells (see diagram). This causes pain and damage to the organ.

The frequency, severity, and duration of the crises vary considerably.



The "Spleen Crisis":

This is painful enlargement of the spleen. 'Sickled" blood accumulates into the spleen, making it swollen, tender and very painful. It may also lead to a **fatal** drop of blood volume from other parts of your body, especially from the brain.

How would I recognize a crisis?

Signs and symptoms of a sickle-cell crises vary from one persons to another. However, common symptoms include:

- Sudden abdominal or chest pain
- Bone pain
- Breathlessness
- Fatigue
- Paleness
- Yellowing of the eyes and skin

How is Sickle-cell Anemia managed?

Children born with sickle-cell disease needs close observation by a pediatrician. But children in a crisis usually need medical attention and a hospitalization may be indicated.

For Pain

Pain management is one of the highest priorities until the crisis stops. With the pediatrician's assessment, strong pain medications; usually given through an IV line or as an injection, is prescribed.

For milder crisis, common pain medications are given instead.

Warmth

Cold air and water can start a painful crisis. Keeping the body warm, especially during cold, moist days can prevent a crisis from happening.

It is a good advice to avoid cold showers, pools or the winter season whenever possible.

Oxygen Therapy