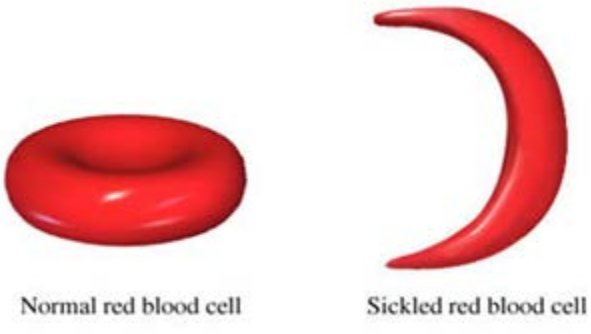
Module 1: Understanding Sickle Cell

Contents

What is Sickle Cell Disease?

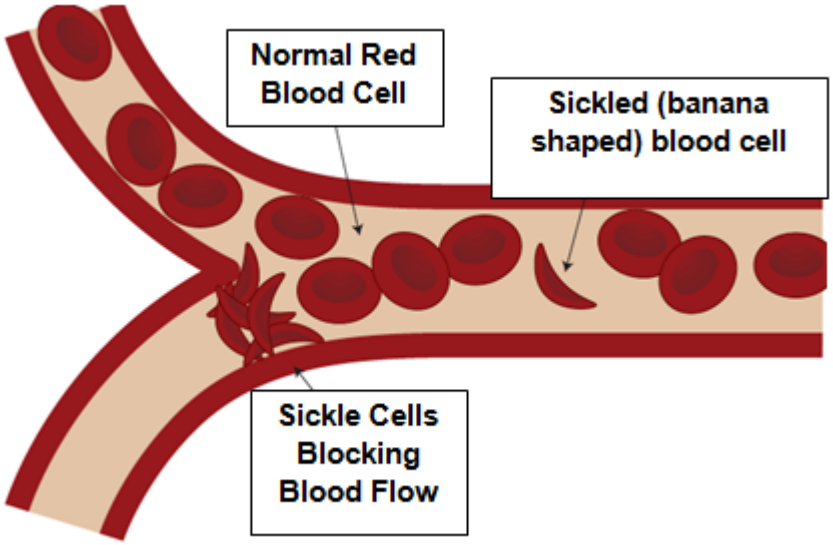
Your body does a lot every day to keep you healthy. One thing your body does is use red blood cells to carry oxygen to different muscles, organs, and tissues. Normal red blood cells are soft and round, like a donut, and travel through the body without any problems.

If you have Sickle Cell Disease, red blood cells can become hard, sticky, and curved in shape, like a banana. Sickled red blood cells are not as soft and flexible as normal cells so they can get stuck in small blood vessels. If sickled red blood cells get stuck in blood vessels, they create an obstacle and block the blood vessel. Then some parts of the body do not get enough oxygen. Not getting enough oxygen throughout the body can cause some pain or damage.



Normal red blood cell Sickled red blood cell

Sickled red blood cells live less time than healthy red blood cells. Sickle red blood cells live only 10-20 days while healthy ones live 120! This means, your body is always short of red blood cells, which can cause you other problems.



What Causes Sickle Cell?

Having Sickle Cell Disease can be a scary thing in your life but knowing the cause of it can help you understand the disease better.

Sickle Cell Disease is an inherited disease which means that it is passed from parents to their children through their genes.

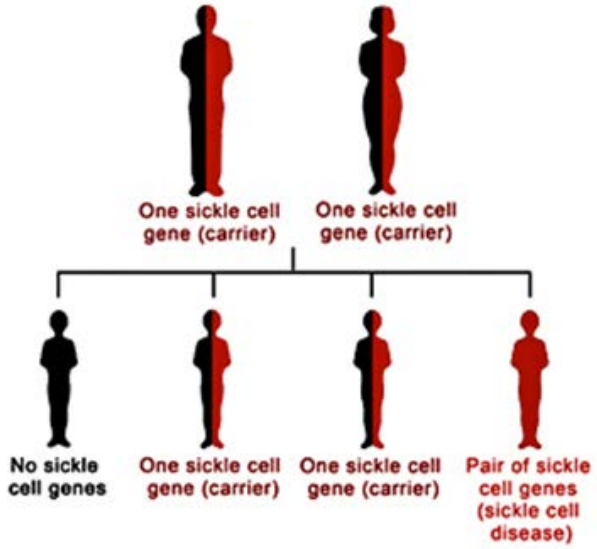
Genes are our body's map for development. We have pairs of genes for the color of our eyes, for our height, for our blood type and for each of our other features, including our hemoglobin type.

Hemoglobin is a protein in red blood cells. Some people have a hemoglobin type that causes red blood cells to be different than normal ones. If parents have this gene, they can pass it onto their children.

Each person has two genes for hemoglobin. You get one gene from your father and one gene from your mother. Sometimes people have just one gene with the different hemoglobin and not the other one. This is called having the trait, but not the disease.

You actually can't know whether the normal hemoglobin gene or the one that is different will be passed on to a child. It is based on chance, like tossing a coin and getting heads or tails. You don't know what you'll get until it happens.

Having sickle cell disease means that both your genes have the unusual hemoglobin types that caused you to have different shaped red blood cells.



More About What Causes It

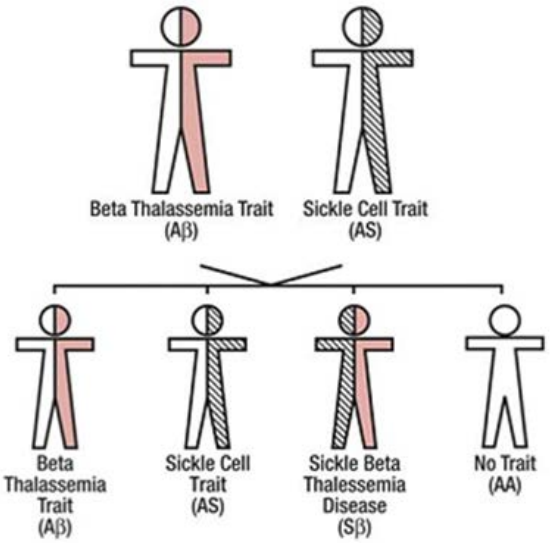
Myth: It is always necessary for both parents to have sickle trait in order to have a child with sickle cell disease.

Reality: If one parent has sickle hemoglobin trait and the other has another abnormal hemoglobin trait, some of their children can also be born with other types of sickle cell disease.

Types of Sickle Cell Disease

Not everyone who has sickle cell disease has the same type. Different gene pairs create a different type of sickle cell. There are different types of unusual hemoglobin (S, C, D, E). To help you understand how sickle cell affects your body. Talk to your doctor to find out yours.

- Hemoglobin SS (Sickle Cell Anemia) the most common type of sickle cell disease with one sickle hemoglobin gene from each parent.
- Hemoglobin SC sickle gene (hemoglobin S) from one parent, hemoglobin C gene from other parent.
- Hemoglobin S beta-plus thalassemia sickle gene from one parent, thalassemia gene from other parent. Thalassemia is a disorder that actually means the body has trouble creating hemoglobin. T
- Hemoglobin S beta-zero thalassemia sickle gene from one parent, beta-zero thalassemia gene from other parent. The beta-zero thalassemia gene makes no hemoglobin.
- Hemoglobin SD-Punjab = sickle gene from one parent, hemoglobin D gene from other parent.
- Hemoglobin SE sickle gene from one parent, hemoglobin E gene from other parent.
- Hemoglobin SO-Arab sickle gene from one parent, hemoglobin O-Arab gene from other parent.



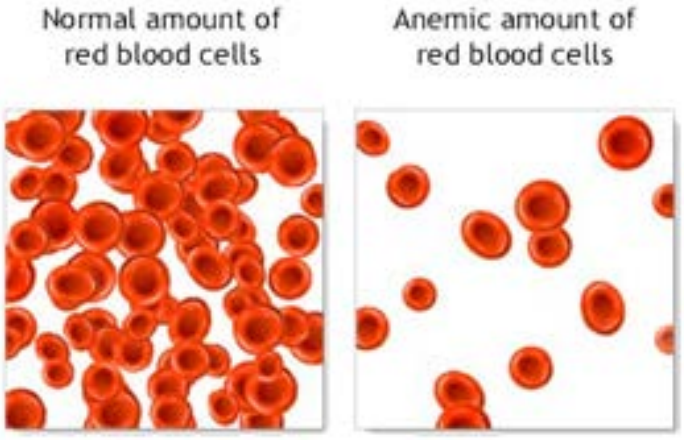
Commons Characteristics Part 1

Having Sickle Cell Disease can mean complications in other areas of your health; read below to learn some characteristics of SCD.

Anemia: Having anemia means that you will have a lower amount of red blood cells and have a lower hemoglobin level. Being anemic can cause weakness and tiring which means you need to rest more often. You may also have to deal with a lot of other symptoms because of your anemia like paleness, coldness or yellowing of the skin, feeling dizzy or even fainting.

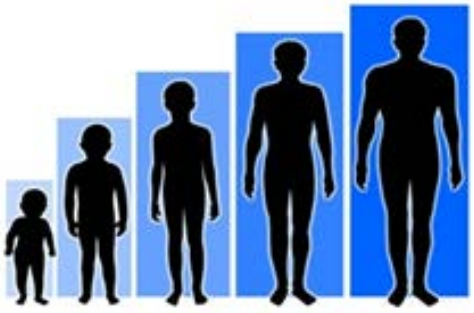
Infection: People with sickle cell disease (especially sickle cell anemia and sickle beta zero thalassemia) are at higher risk of infection than people who do not have sickle cell disease. It is very important for infections to be treated quickly. Some common infections for people with sickle cell disease include:

- Meningitis - infection of the spinal fluid
- Pneumonia - infection of the lungs
- Osteomyelitis - infection of the bones
- Sepsis - infection in the blood



Common Characteristics Part 2

Growth and Development: Children and younger patients with Sickle Cell Disease (SCD) may be smaller and thinner than people their age who do not have SCD. Because of the anemia associated with SCD, their metabolism can increase causing them to burn more calories and put on less weight. Even though they may be smaller than average, many children are able to "catch up" in their teenage or early adult years.



Decreased Urine Concentration: Urine concentration is the gathering of waste from the body in the urine so that it can be removed when you use the restroom. Sickle cell disease may cause you to urinate often because your body is not able to concentrate or collect waster in your urine. The sickle cells in your blood can cause them to block blood flowing through your blood vessels and this blockage can actually harm your kidneys. Your kidneys help to collect the waste in your body so if they don't work right, then that affects your urine. You actually should be drinking a lot of water to help the blood flow and so you may have to go to the restroom a lot.

Jaundice/Yellow Eyes: People with sickle cell disease sometimes seem to have a yellow color over their eyes. This has to do with what is happening with your red blood cells. Red blood cells in the body break down quickly. When this happens, bilirubin, an orange-yellow color is released from the liver. If too much bilirubin fills the body, it causes jaundice, which is when the white part of the eye or your skin turns yellow. The yellow color may come and go but some people may always have yellow eyes. You can't pass this on to other people and you do not need to get medical help for it.

Where to Find More Information

If you want to read more about any of the topics in this module, here are the website references where you can find more information:

The Sickle Cell Disease Association of America is an organization that offers information and support to people and families dealing with sickle cell.

St. Jude created a guide just for teens! It reviews a lot of the same information and gives you a different perspective.

Kidshealth is great website that explains a lot of the information shared in this module.

Great job!!

You're one step closer to finishing the CHECK Sickle Cell Online Program. Hopefully you now know more about sickle cell and how it affects your body.

Now you can move onto the next topic and learn all about different sickle cell treatments!