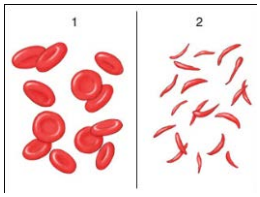
Sickle Cell Disease – The Basics

What is Sickle Cell Disease?

Sickle cell disease is an inherited disorder affecting a protein in the red blood cells called *hemoglobin*. Hemoglobin is responsible for carrying oxygen throughout the body. Sickle cell disease occurs when there are genetic mutations in the hemoglobin. While normal red blood cells contain a molecule called *hemoglobin A,* sickle red blood cells contain an abnormal version called *hemoglobin S* ("S" is for sickle).

Cells made with normal hemoglobin are fluid, round, and shaped like a doughnut without a hole. Sickle cells start out this same shape and are able to pick up oxygen normally. However, when the oxygen is dropped off at the body's tissues, a chemical reaction within the cell forms rigid rods that change the red blood cell into a sickle or crescent-moon shape.



1. Normal red blood cells
2. Sickle cells

These sickle-shaped cells are rigid and sticky. They get trapped within the blood vessels and interfere with normal blood flow. These blockages can occur anywhere in the body and deprive tissues and organs of oxygen. This causes tissue damage, which then can lead to pain and organ dysfunction.

Sickle cells have a much shorter life span (10 to 20 days) than normal red blood cells (90 to 120 days). Even though our bodies produce new red blood cells every day, people with sickle cell anemia cannot make enough to keep up normal levels or red blood cells. This constantly lowered red blood cell count is called *anemia.*

Sickle cell disease is a serious, complicated disease. It is very important that people with sickle cell disease take good care of themselves, attend their medical appointments, and seek treatment as soon as complications arise. With proper comprehensive care, people with sickle cell disease can live long, productive lives.

Where can I find more information?

Kid's Health – Information for Parents: http://kidshealth.org/parent/medical/heart/sickle_cell_anemia.html

Kid's Health – Information for Kids: http://kidshealth.org/kid/health_problems/blood/sickle_cell.html

The Sickle Cell Information Center: www.scinfo.org

National Heart, Blood and Lung Institute: http://www.nhlbi.nih.gov/health/dci/Diseases/Sca/SCA_WhatIs.html

St. Jude's Hospital: http://www.stjude.org/stjude/v/index.jsp?vgnextoid=0f3c061585f70110VgnVCM1000001e0215acRCRD

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Revised May 2019