

Sickle C Disease (Hb S/C)

What is Hb S/C?

Hemoglobin Sickle C Disease (Hb S/C) is a “mild” form of sickle cell anemia. The red blood cells (RBCs) of a child with Hb S/C disease have two kinds of abnormal hemoglobin. They are called hemoglobin S and hemoglobin C.

What Causes Hb S/C?

The hemoglobin in the RBCs carries oxygen to all parts of the body. Regular RBCs are round like a donut. The RBCs in a child with Hb S/C disease are misshapen. Some even look like the sickle-shaped cells found in sickle cell anemia. Others are folded or football-shaped.

What Symptoms or Problems Occur with Hb S/C?

(Symptoms are something out of the ordinary that a parent notices.)

Anemia – Your child will always have a slightly low red blood count. This is called anemia. This mild anemia is usually no problem, but sometimes causes tiredness and/or weakness.

Pain – The red blood cells of Hb S/C disease are rigid and stiff. Sometimes they “clog up” the small vessels in the bones and other parts of the body. This can cause pain because enough oxygen cannot get into the bones. The pain usually happens in the arms, legs, stomach, and/or back. It can last for hours, for days, or up to a week. It can vary from mild to moderate to severe. The location, length, and amount of pain can vary. How often this happens also varies. Some children with Hb S/C disease have no periods of pain at all, but most will have a few each year.

Pneumonia and Other Infections – A child with Hb S/C disease has a higher chance of getting certain infections, especially pneumonia. The abnormal RBCs can “clog up” in the lungs and cause infection. This is called “chest syndrome.” Problems to watch for include fever, fast breathing, trouble breathing, retractions (ribs “suck in” when breathing), very congested cough, and chest pain. Your child should see a doctor immediately if these problems happen.

Spleen – The spleen is a small organ located on the upper left side of the stomach area, up under the rib cage. It helps fight infection in the body. Children with Hb S/C disease may have an enlarged (big) spleen. This doesn’t happen until they are about five years or older. A big spleen usually doesn’t cause any problems. Sometimes teen-agers and/or adults can have pain near the spleen and a drop in the red blood count. This is called a “spleen crisis.”

Eyes – Older children (over age ten) and adults with Hb S/C disease may develop damage to the retina in the back of the eye. This can cause blindness if not treated early. Regular eye checkups by a medical eye doctor (ophthalmologist) are needed to diagnose and treat this problem.

What is the Treatment for Hb S/C?

Medication – The infant or young child with Hb S/C disease is more likely to have infection of the blood (septicemia). Scientific studies show that penicillin can prevent death from septicemia. When taken every 12 hours, penicillin can kill bacteria before they grow in the blood and cause septicemia.

Fluids – The infant or young child with Hb S/C disease needs to drink plenty of liquids to keep blood vessels open.

Things to Remember

Children with Hb S/C disease can experience periods of pain. Medications for pain, such as acetaminophen (Tylenol) or ibuprofen (Advil) often help the pain. Sometimes children won’t use the part of the body that hurts, even when it feels better. Don’t force your child to stand or walk. Children will be active again when they feel like it. Rest and drinking plenty of liquid can help the pain. A heating pad can also help. If a child’s pain isn’t better after taking medicine at home, he or she may need to be treated with a stronger medicine in the hospital.