

Sickle-Beta+-Thalassemia Fact Sheet

What is sickle-beta+-thalassemia?

Sickle-beta+-thalassemia is a milder form of sickle cell anemia. Hemoglobin is the part of the red blood cells that carries oxygen throughout the body. The most common (“normal”) form is called hemoglobin A. Your child’s red blood cells definitely contain some sickle (S) hemoglobin, but may also display the hemoglobin disorder called beta+ thalassemia.

How does someone get sickle-beta+-thalassemia?

Sickle-beta+-thalassemia is an inherited condition. One parent carries sickle trait, which causes no health problems for him or her. The other carries the trait for beta+ thalassemia, which sometimes causes a mild anemia (low blood count), but usually no symptoms. When these traits come together in the same person, the result is sickle-beta+-thalassemia. *When one parent has sickle trait and the other has beta+ thalassemia trait, each baby they have together has a 1 in 4 chance of inheriting sickle-beta+-thalassemia.*

How is sickle-beta+-thalassemia diagnosed?

The screening test performed by the Health Department merely suggests the possibility of sickle-beta+-thalassemia. To help confirm the presence of this disorder, your doctor should perform a test called an *electrophoresis* on your baby at around 2-3 months of age. This test involves taking a small amount of blood from the baby and sending it to the laboratory. In some cases, the test will show that the baby simply has sickle cell trait, for which no additional follow-up or treatment is needed.

What problems can sickle-beta+ thalassemia cause?

In people confirmed to have sickle-beta+-thalassemia, symptoms and problems range from very mild to somewhat severe. This is because with beta+ thalassemia, production of hemoglobin A ranges from fairly high in some people, to very low in others. In affected people who produce a lot of hemoglobin A, symptoms are mild. In those who produce very little hemoglobin A, problems and symptoms can be severe and similar to those of classic sickle cell anemia. Such problems include moderate to severe anemia (low blood count); episodes of pain in the arms, legs, stomach, and back (*pain crises*); increased risk of serious (blood) infections in younger children; gallstones; enlarged spleen; and risk of sudden drops in blood count, particularly with certain infections (*aplastic crises*).

Because the newborn screening test does not tell for certain whether your baby has sickle-beta+-thalassemia, it is important to have the electrophoresis test done at 2-3 months of age. Due to the possible increased risk of serious infections in younger children with the disorder, your doctor may choose to start your baby on oral penicillin before the test is performed. If the test does confirm sickle-beta+-thalassemia, your baby will need to have regular check-ups with a knowledgeable physician. In some cases, consultation with a blood specialist might also be recommended.