

Sickle Beta Zero Thalassemia (S β^0 -Thalassemia)

What is S β^0 -Thalassemia?

Hemoglobin in the red blood cells carries oxygen to all parts of the body. With Sickle Beta Zero Thalassemia (S β^0 -Thalassemia), the red blood cells have abnormal hemoglobin. This is called hemoglobin S (sickle hemoglobin). Also, there is a problem called thalassemia. This causes red blood cells to be small and pale.

What Causes S β^0 -Thalassemia?

S β^0 -Thalassemia is an inherited disease. Hemoglobin S is inherited from one parent. Thalassemia is inherited from the other parent. In S β^0 -Thalassemia the red blood cells are not round. They are misshapen. Some may have a sickle (or banana) shape.

What Symptoms or Problems Occur with S β^0 -Thalassemia?

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

The symptoms of S β^0 -Thalassemia depend on the type and seriousness of the disease. Problems happen when different parts of the body don't get enough oxygen. This occurs when there is low hemoglobin and not enough red blood cells in the blood (anemia).

Problems include:

- ◆ feeling tired and weak
- ◆ slightly enlarged spleen (an organ located on the upper left side of the stomach area)
- ◆ mild anemia
- ◆ periods of pain
- ◆ increased risk for infections

What is the Treatment for S β^0 -Thalassemia?

Medications – Your child has more risk of getting infections. These can be infection in the brain, pneumonia, blood poisoning, and bone infection. Your child's doctor will probably give him or her penicillin to help prevent infections. It is important that your child take the penicillin as directed. Any fever of 101°F (38.5°C) is serious. Take your child to the doctor or clinic IMMEDIATELY if this happens.

Periods of pain can happen with S β^0 -Thalassemia. The red blood cells are rigid and stiff. Sometimes they “clog up” the small blood vessels in the bones and other parts of the body. This can cause pain because not enough oxygen can get into the bones and other parts of the body. The pain usually happens in the back, stomach, arms, and legs. There may be swelling. Pain can last for a few hours or up to a week or more. The amount of pain varies. Medicines help ease the pain.

Sometimes children with S β^0 -Thalassemia have a slightly enlarged spleen. A bigger spleen usually doesn't cause any problems.

Things to Remember

Children with S β^0 -Thalassemia can have normal lives and life spans. Don't think of your child as “sick.” You should treat him or her normally. Your child will need to see the doctor for regular checkups and vaccinations. He or she will also need to make several visits a year to see a hematologist (a doctor who is a blood specialist).