Sickle Beta Zero Thalassemia Disease

WHAT IS SICKLE BETA THALASSEMIA ZERO (Hb Sβ0 thal)

Sickle Beta Zero Thalassemia (Sickle BA-ta zero thal-a-SEE-me-a) is similar to sickle cell anemia. Your child's red blood cells contain abnormal hemoglobin, called "hemoglobin S" or "sickle hemoglobin". In addition, the red blood cells have a defect called thalassemia, which results in cells that are small in size and more pale than usual.

Instead of appearing round or donut shaped, your child's red blood cells are somewhat small, pale, and misshapen. Some may appear sickled or banana shaped.

Because sickle beta zero thalassemia is inherited, it is a lifelong disorder. There is no treatment or cure. Your child will always have a mild anemia or slightly low blood count. This may result in occasional tiredness or weakness.

HOW DID MY CHILD GET SICKLE BETA ZERO THALASSEMIA

When one parent has Sickle Trait (AS) and the other parent has Beta Zero Thalassemia Trait (AT) there is a 1-in-4 chance (25 percent) their baby will have normal hemoglobin (AA), Beta Thalassemia Zero Trait (AT), Sickle Beta Zero Thalassemia (STO), or Sickle Trait (AS). These chances remain the same for each pregnancy.
Most information about sickle cell anemia also applies to sickle beta zero thalassemia. Like sickle cell anemia, your child has an increased risk of getting infections, such as meningitis (an infection of the brain), pneumonia, septicemia (blood poisoning), and osteomyelitis (an infection of the bone). Because your child is more susceptible to infections than other children, he or she will be put on penicillin until age five in order to help prevent these infections from occurring. It is important that your child take penicillin as directed. Any fever of 101.5°F or 38.5°C should be taken seriously and your child taken to your family doctor or clinic IMMEDIATELY!

Painful episodes can occur with sickle beta zero thalassemia. The sickled red blood cells in sickle beta zero thalassemia, like those in sickle cell anemia, are rigid and stiff and may sometimes cause "log jams" in the small blood vessels in the bones, organs and other parts of the body. Since oxygen (which is carried by the red blood cells) cannot get past the "log jams" into the bones and organs, this can cause pain. These painful episodes occur most commonly in the back, stomach, arms, and legs. They can last for a few hours to as much as a week or more. Occasionally, the pain is accompanied by swelling. Painful episodes can vary in severity of the pain and the duration. There are medications to help ease the pain and make your child more comfortable.

Often, children with sickle beta zero thalassemia have a slightly enlarged spleen, but this usually does not cause any problems.

**IT IS IMPORTANT TO INFORM ANY PHYSICIAN THAT SEES YOUR CHILD THAT HE OR SHE HAS SICKLE BETA ZERO THALASSEMIA!**

**WHAT YOU CAN EXPECT**

A child with sickle beta zero thalassemia can have a normal life-style and life span. Your child should not be considered "sick", and you should treat him or her normally. He/She will need to be seen regularly by your family doctor for regular check-ups and vaccinations. Your child will also need to make several visits a year to see a hematologist or blood specialist.