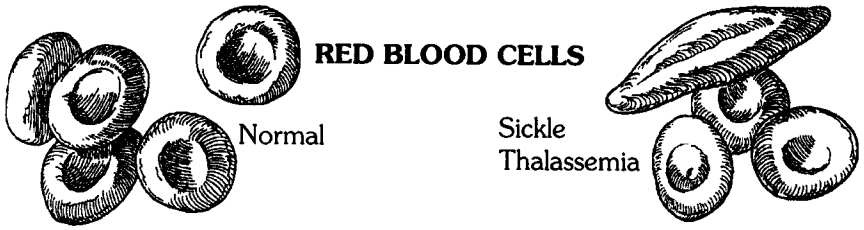
NEPSCC
New England Pediatric Sickle Cell Consortium

Sickle Beta Plus Thalassemia Disease

WHAT IS SICKLE BETA PLUS THALASSEMIA (Hb Sβ$^+$ thal)

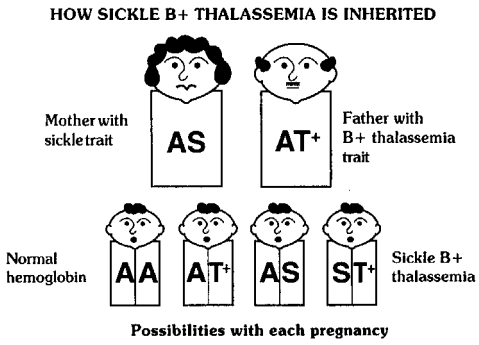
Sickle Beta + Thalassemia (Sickle BA-ta Plus Thal-a-SEE-me-a) is a "mild" form of sickle cell disease. Your child's red blood cells contain an abnormal hemoglobin called hemoglobin S or sickle hemoglobin in addition to a small amount of the normal hemoglobin called hemoglobin A. The red blood cells have a defect called beta plus thalassemia, which results in cells which 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.



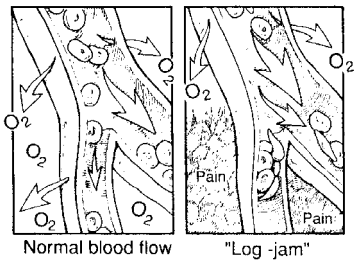
Because sickle beta plus thalassemia is inherited, it is a lifelong disorder. There is no treatment or cure. Your child will always have a mild anemia or slightly low blood count. This may cause occasional tiredness or weakness.

HOW DID MY CHILD GET SICKLE BETA PLUS THALASSEMIA



When one parent has Sickle Trait (AS) and the other parent has Beta Thalassemia Plus Trait (AT+), them is a l-in-4 chance (25 percent) the baby will have normal hemoglobin (AA), Sickle Trait (AS), Beta Thalassemia Plus Trait (AT+), or Sickle Beta Plus Thalassemia (ST +). These chances remain the same for each pregnancy.

PROBLEMS SEEN IN CHILDREN WITH SICKLE BETA PLUS THALASSEMIA



Painful episodes can occur with sickle-thalassemia. The sickled red blood cells in sickle-thalassemia, somewhat 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 medicines to help ease the pain and make your child more comfortable.

Your child has an increased risk of getting infections, especially pneumonia. The abnormal sickled red blood cells "clog up" the lungs and thus increase the risk of infection there.

Sometimes children with Sickle Beta Plus 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 PLUS THALASSEMIA!

WHAT YOU CAN EXPECT

A child with sickle beta plus 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.

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