Sickle-Hemoglobin E Disease Fact Sheet

What is sickle-hemoglobin E disease?

Sickle-hemoglobin E disease is a milder form of sickle cell anemia. Hemoglobin is the part of the red blood cells that carries oxygen through the body. The most common ("normal") form is called hemoglobin A. Your child's red blood cells contain two different hemoglobins, called hemoglobin S and hemoglobin E.

How does someone get sickle-hemoglobin E disease?

Sickle-hemoglobin E disease is an inherited condition. One parent contributes the sickle (S) hemoglobin, and the other contributes the E hemoglobin. Sickle hemoglobin is found most frequently among people of African or Mediterranean descent. Hemoglobin E occurs most often among people of Southeast Asian origin. For each parent, the respective traits cause no health problems. When together in the same person, however, the result is sickle-hemoglobin E disease. *When one parent has sickle trait and the other has hemoglobin E trait, each baby they have together has a 1 in 4 chance of inheriting sickle-hemoglobin E disease*.

What problems can sickle-hemoglobin E disease cause?

Most people with sickle-hemoglobin E disease have mild to moderate anemia (low blood count). Although occasionally causing some tiredness or weakness, usually this low blood count does not cause serious symptoms or problems. Many of the red blood cells in people with sickle-hemoglobin E disease are "stickier" than usual, and thus may "clog up" the small blood vessels in the bones and other parts of the body. When this happens, the person will often experience pain in the bones or other body tissues. Painful episodes most often affect the arms, legs, stomach, and back. Such episodes may last for hours, days, or even a week. The amount of pain in each episode also varies, ranging from mild to severe.

Children with sickle-hemoglobin E disease often have an increased risk of getting certain infections, especially pneumonia. In the presence of pneumonia, the abnormal red blood cells can also "clog up" small blood vessels in the lungs, causing *acute chest syndrome*. Symptoms to watch for include fever, fast breathing, trouble breathing, retractions (spaces between the ribs suck in when breathing), very congested cough, and chest pain. If these symptoms occur, your child should see a doctor immediately.

The spleen in younger children with sickle-hemoglobin E may be somewhat less effective in removing bacteria (germs) from the blood. This can lead to a slightly higher risk of serious (blood) infections in younger children with the disorder. The spleen lies under the rib cage in the upper left part of the abdomen. It is often enlarged in children with sickle-hemoglobin E disease, especially as they get older. The enlarged spleen itself usually does not cause any problems. However, teenagers and adults with sickle-E disease may have an episode of pain over the spleen and sudden drop in blood count, called a *splenic crisis*.

Are there any special treatments for people with sickle-hemoglobin E disease?

Because of the slightly higher risk of serious infections in younger children, most blood specialists recommend treatment with oral penicillin starting in the first two months of life, and continuing through five years of age. This treatment is intended to prevent serious blood infections. All routine vaccinations and other recommended immunizations should be taken on time. *Children with sickle-hemoglobin E disease should be seen regularly by a knowledgeable physician and/or blood specialist*. Because of the risk of damage over time to the retina (in the back of the eye), older children with sickle-hemoglobin E should also be seen regularly by an ophthalmologist (medical eye doctor).

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