Sickle Cell Anemia & Hemoglobin C

MISSOURI DEPARTMENT OF HEALTH AND SENIOR SERVICES
Missouri Sickle Cell Anemia Program

MISSOURI DEPARTMENT OF HEALTH AND SENIOR SERVICES
HEMOGLOBIN

Hemoglobin is that portion of the red blood cells which carries oxygen to all parts of the body. The most common type of hemoglobin is called Hemoglobin A. Sometimes there are changes in Hemoglobin A, and other types of hemoglobin are formed.

Genes are units of hereditary material which are passed from parents to their children. These genes, which act in pairs, determine the physical characteristics unique to each individual. One gene in each pair is contributed by the mother and the other gene in the pair is contributed by the father. Just as genes for eye color, hair color, and many other traits are inherited, so are genes for hemoglobin. If a person inherits a Hemoglobin A gene from each parent (AA), that person will have normal hemoglobin.

This discussion is related to two of the possible changes in normal hemoglobin: Hemoglobin S and Hemoglobin C.

HEMOGLOBINS

Sickle Cell Anemia

If a person inherits a gene for Hemoglobin S from both parents (SS), that person will have sickle cell anemia. In sickle cell anemia the hemoglobin releases too much oxygen, causing the normally round blood cells to be sickle-shaped. These sickle cells tend to die more quickly than normal red blood cells and the body doesn't receive enough oxygen. This may cause the person to be pale, short of breath and easily tired. At times, the sickle cells may become stuck in the small blood vessels and cause pain and damage to major organs. This condition is known as "sickle cell crisis." Other medical problems include slowed growth, more frequent infections, jaundice and blood in the urine. These symptoms and their severity differ from person to person. Sickle cell anemia affects one out of every 400-600 black Americans. Although it is most common in blacks, it also occurs in people of Mediterranean, Middle Eastern, East Indian, Latin American and Asian descent.

Sickle Cell Trait

Sometimes a person will inherit a gene for normal hemoglobin from one parent and a gene for Hemoglobin S from the other parent. This is Hemoglobin AS, also known as sickle cell trait. Sickle cell trait occurs in one out of every 10-12 black Americans and usually causes no health problems. Sickle cell trait is not a mild form of sickle cell anemia. However, there is reason for concern. When both parents have sickle cell trait (AS), their children may inherit two genes for sickle hemoglobin and have sickle cell anemia.
When both parents have sickle cell trait:

- The chances with each pregnancy are one in four that a child will have normal hemoglobin (AA); two in four that a child will have sickle cell trait (AS); and one in four that a child will have sickle cell anemia (SS).

When one parent has normal hemoglobin and the other parent has sickle cell trait:

- The chances with each pregnancy are two in four that the child will have normal hemoglobin (AA) and two in four that the child will have sickle cell trait (AS). There is not chance that the child will have sickle cell anemia.

**BOTH PARENTS MUST HAVE AT LEAST ONE GENE FOR HEMOGLOBIN S TO HAVE A CHILD WITH SICKLE CELL ANEMIA.**
HEMOGLOBIN C

Hemoglobin C Disease

If a person inherits a gene for Hemoglobin C from both parents (CC), that person will have Hemoglobin C disease. This disease may produce mild to moderate anemia, jaundice, enlarged spleen and gallstones. However, some people with Hemoglobin C disease do not have any of these symptoms. Hemoglobin C disease is relatively rare and appears in approximately one out of every 10,000 black Americans.

in every 10,000

Hemoglobin C Trait

A person who inherits a Hemoglobin A gene from one parent and a Hemoglobin C gene from the other parent, will have Hemoglobin C trait (AC). Approximately one in every 50 black Americans has Hemoglobin C trait. This is not a disease and there are no health problems associated with it.

in every 50

When both parents have Hemoglobin C Trait:

the chances with each pregnancy are one in four that a child will have normal hemoglobin (AA); two in four that the child will have hemoglobin C trait (AC); and one in four that the child will have Hemoglobin C Disease (CC).
SICKLE CELL HEMOGLOBIN C DISEASE

If a person inherits a gene for Hemoglobin S from one parent and a gene for Hemoglobin C from the other parent, that person will have sickle cell Hemoglobin C disease (SC disease).

This disease may produce symptoms similar to sickle cell anemia or Hemoglobin C disease. Some cases may be mild and have few symptoms while others may be more severe and have many symptoms. While symptoms of this disease usually appear during the first ten years of life, they may not appear until later. Sometimes they first appear when a woman becomes pregnant. These women may experience a higher rate of complications during pregnancy. SC disease is considered to be less severe than sickle cell anemia; however, damage to hip joints and the retina are more common in persons with SC disease. Approximately one out of every 1,000 black Americans has SC disease.

The chances with each pregnancy are one in four that a child will have normal hemoglobin (AA); one in four that a child will have hemoglobin C trait (AC); one in four that a child will have sickle cell trait (AS); and one in four that a child will have sickle cell hemoglobin C disease (SC).
**What Are The Most Common Hemoglobin Types?**

<table>
<thead>
<tr>
<th>Hemoglobin Type</th>
<th>Condition</th>
<th>Symptoms</th>
<th>Inheritance</th>
</tr>
</thead>
<tbody>
<tr>
<td>*AA</td>
<td>Most common type of hemoglobin</td>
<td>None</td>
<td>Will always pass on a normal hemoglobin gene to offspring</td>
</tr>
<tr>
<td>*AS</td>
<td>Sickle cell trait</td>
<td>None</td>
<td>50% chance of passing on a sickle cell gene to offspring</td>
</tr>
<tr>
<td>SS</td>
<td>Sickle cell anemia</td>
<td>Varies among individuals and may include paleness, shortness of breath, easily tired, pain, slowed growth, frequent infections, jaundice, and blood in the urine</td>
<td>Will always pass on a sickle gene to offspring</td>
</tr>
<tr>
<td>*AC</td>
<td>Hemoglobin C trait</td>
<td>None</td>
<td>50% chance of passing on a hemoglobin C gene to offspring</td>
</tr>
<tr>
<td>CC</td>
<td>Hemoglobin C disease</td>
<td>May or may not have anemia, jaundice, enlarged spleen, or gallstones</td>
<td>Will always pass on hemoglobin C gene to offspring</td>
</tr>
<tr>
<td>SC</td>
<td>Sickle cell hemoglobin C disease</td>
<td>May be similar to sickle cell anemia or hemoglobin C disease. May have complications during pregnancy</td>
<td>Will always pass on either a sickle gene or a hemoglobin C gene to offspring</td>
</tr>
</tbody>
</table>

*As you can see, if one gene in the pair is Hemoglobin A, there will usually be no health problems associated with that condition.
IMPORTANCE OF KNOWING YOUR HEMOGLOBIN TYPE

— Hemoglobin abnormalities are a major health problem in the United States.

— You will not know if you have a gene for an unusual type of hemoglobin unless you are tested.

— If you have an unusual hemoglobin trait, it is possible for you to pass it on to your children.

— If one parent is found to have an unusual hemoglobin trait, the other parent may wish to be tested to determine whether the couple might have an affected child.

— If both parents have an unusual hemoglobin type, counseling will identify their chance of having a child with abnormal hemoglobin.

— It is advisable to test children of people who have unusual hemoglobin.

— Unusual hemoglobins are not contagious and do not affect intelligence.

— As yet there is no cure for the conditions associated with unusual hemoglobins, but treatment is available.

For additional information and testing services, contact your family physician, local health department or

MISSOURI DEPARTMENT OF HEALTH
AND SENIOR SERVICES
P.O. Box 570
Jefferson City, MO 65102-0570
1-800-877-6246