

So, What is Hemoglobin H Disease?

People with hemoglobin H disease have only 1 working gene to make alpha globin. As a result, they make less than the usual amount of alpha globin. This leads to fewer red blood cells, also known as anemia.

Many people with hemoglobin H disease do not have serious health problems. However, since they have fewer red blood cells, they often have mild to moderate anemia, which can cause them to feel more tired. Sometimes the anemia can get worse or lead to other problems.

High fevers can cause the anemia to worsen. Also, there are some medications and chemicals that can cause patients to become more anemic. Some of these medicines and household products are listed in this brochure. Your doctor or counselor can also tell you which medicines and other chemicals to avoid.

What is Hemoglobin H – Constant Spring Disease?

Hemoglobin H – Constant Spring is a disease that is caused by an even more unstable alpha globin. This causes the red blood cells to break down faster than usual so there are even fewer red blood cells in the body. This leads to more severe anemia. Other problems often include a large spleen, gallstones, an increased risk for infections, jaundice, and leg ulcers. These people may also need to have blood transfusions either occasionally, or regularly.



What is Hemoglobin H Disease?

In order to understand hemoglobin H, it helps to understand a little more about our blood. Hemoglobin H is a form of alpha thalassemia, an inherited blood disease that affects a part of the blood called hemoglobin.

Hemoglobin

One role of the blood is to take oxygen from air breathed into the lungs and bring it to the rest of the body. The part of the blood that does this is the red blood cell. Hemoglobin is the part of the red blood cell that carries the oxygen.

The type of hemoglobin that is made depends on the genes the child inherits from each parent. Genes carry instructions on how to make different parts of a person. Every gene carries different instructions.

We inherit our genes from our parents in pairs. One half of each pair comes from each parent. In this way a child is made up of instructions from both mom and dad in an equal amount. Each child inherits genes from the father and genes from the mother that instructs the body how to make hemoglobin.

There are two kinds of protein found in the normal adult hemoglobin: alpha globin and beta globin. The normal adult hemoglobin has 2 alpha globins and 2 beta globins. The amount of alpha globin depends on the number of working genes. Most people have four working genes that make alpha globin.

What is Alpha Thalassemia?

There are several forms of alpha thalassemia. These different types result from the number of working alpha globin genes. The fewer working genes a person has, the more serious his or her type of alpha thalassemia will be. People with 3 working genes are called silent carriers. Most of these people will never know they have 1 non-working gene. People with 2 working genes have alpha thalassemia trait. People with alpha thalassemia trait do not have any health problems and may never know they have only 2 working genes. This is because these 2 genes make enough alpha globin protein to keep the person healthy. People with only 1 working gene have hemoglobin H disease. There are also people who have no working genes. They have alpha thalassemia major.



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What is Hemoglobin H?

Alpha thalassemia is very common in people with ancestors from China, the Philippines, Thailand, Vietnam, Cambodia, Laos, and other Asian countries. It is also common in people of African ancestry, including West Indian and African Americans. However, people from any ethnic group can have a type of alpha thalassemia.

How Can I Care for My Baby?

The doctor and staff at the hematology center will discuss how you can best care for your baby. With proper treatment most people with hemoglobin H disease can lead relatively normal lives. However, children with hemoglobin H disease are more likely than other children to get infections. These infections and fevers may cause the red blood cells to break down faster, leading to anemia.

If your baby shows any of these signs of severe anemia, contact your doctor or hematology center immediately.

- Pale or yellowish skin
- Yellow eyes
- Extreme fatigue
- Abdominal or back pain
- Dark black stool
- Dark orange urine

To avoid any of the above problems, call your baby's doctor or the hematology center whenever your child becomes sick.

People who have hemoglobin H disease need to avoid certain medicines, household products, and foods. They may cause anemia to worsen if your child comes in contact with them. Many people with hemoglobin H disease may also take a B vitamin supplement called folic acid. If your child receives blood transfusions, he or she may also develop iron overload. Your child should not be given any iron supplements unless a blood test shows that he or she is iron deficient. Please check with your hematology center before giving your child any iron supplements.



What Should My Child Avoid?

On the following page is a list of medicines and household products to avoid. Take this list to any doctor's or emergency room visit and tell the doctors that your child has hemoglobin H disease. Also show this list to the pharmacist who is filling prescriptions for your child. Do not give your child any over the counter medicines without talking to your doctor first.

MEDICATIONS AND PRODUCTS TO AVOID

SULFA DRUGS

Sulfacetamide (eye drops)
Sulfapyridine
Sulfasalazine
(Salicylazosulfapyridine)
Sulfanilamide
Dapsone

ANTIMALARIALS

Primaquine
Chloroquine
Hydroxychloroquine sulfate

OTHER ANTIBACTERIALS

Nalidixic acid (Negram)
Nitrofurantoin
Furazolidone
Chloramphenicol
Beta-aminosalicylic acid
Ciprofloxacin
Doxycycline

ANALGESICS

Aspirin*
Phenacetin
Acetanilide

TUBERCULOSIS DRUGS

Isoniazid
Rifampin
Folic Acid Antagonists
Pyrimethamine

OTHER ITEMS

Iron Supplements
Vitamin K analogues
Quinidine Gluconate
Phenazopyridine (pyridium)
Toluidine Blue (a dye)
Methylene Blue (a dye)
Naphthalene (Mothballs)
Fava Beans

*(Acetaminophen is safe as an alternative)

Your child should also avoid contact with fava beans and mothballs. Both of these can cause severe anemia. Other types of beans are not a problem. Swallowing or inhaling mothballs can be very harmful to your child. Call your doctor immediately if your child swallows a mothball.

How Do I Find Out If I Am At Risk of Having a Baby with Hemoglobin H Disease?

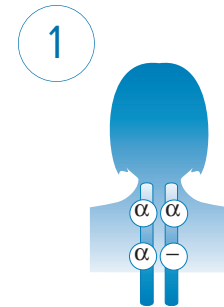
The first step is to find out if you have alpha thalassemia trait. Ask your doctor to look at the size and number of your red blood cells. These are both common readings when you have a Complete Blood Count (CBC).

Your doctor may then need to request more specialized tests, such as a hemoglobin electrophoresis and a blood iron level.

These tests will show your doctor if you have any type of alpha thalassemia trait.

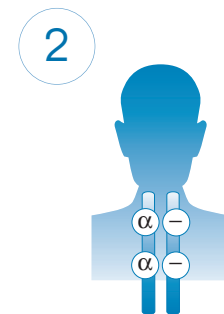
If you have alpha thalassemia trait and are considering having a child or are already pregnant, your partner should be tested to see if he or she has any type of alpha thalassemia trait.

There are two kinds of **alpha thalassemia trait**:



Silent carrier:

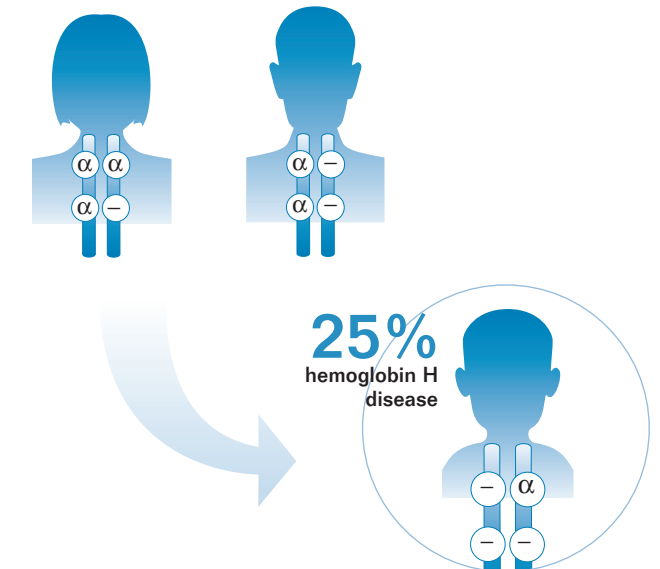
This condition causes no health problems and can only be diagnosed by special DNA testing.



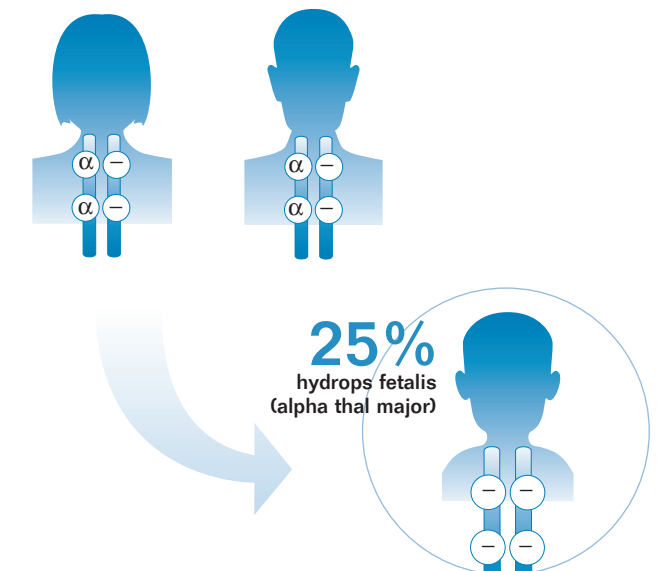
Alpha thalassemia trait:

This condition also generally causes no health problems other than a possible mild anemia. The red blood cells are smaller than usual. Doctors often mistake **alpha thalassemia trait** for iron deficiency anemia.

If one parent has the trait for **alpha thalassemia** and the other parent is a silent carrier, there is a **25% chance with each pregnancy** that their child will be born with **hemoglobin H disease**. This condition can result in serious health problems, such as enlargement of the spleen, bone deformities, and fatigue.



If both parents carry **alpha thalassemia trait**, there is a **25% chance with each pregnancy** that the child may inherit **hydrops fetalis**, also known as **alpha thalassemia major**. This condition requires treatment during pregnancy in order for the baby to survive.



Inform your doctor if you have alpha thalassemia trait. Discuss what it might mean for your unborn child. There are tests that can tell you which genes your unborn child has inherited from you and your partner and what type of alpha thalassemia he or she has. You can discuss these tests with your doctor for more information.



For more information about thalassemia, please contact: Cooley's Anemia Foundation at (800) 522-7222 or info@cooleysanemia.org. You can also visit our web site at www.cooleysanemia.org

If you are tested for **alpha thalassemia trait** and are found to be a trait carrier, your spouse should also be tested for **alpha thalassemia trait**.

If you and your spouse both carry **alpha thalassemia trait** and plan to have children, you should seek the advice of a genetic counselor.