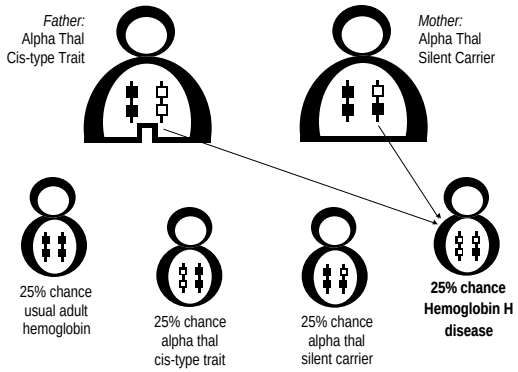
How do you get Hgb H disease?

Hgb H is an inherited disease. It is passed on from a child's mother and father through their genes. This means that the disease is something you have at birth and throughout your life. It is not a communicable disease; you cannot "catch it" from someone else.

Hemoglobin is made of three parts: a heme protein, an alpha globin and a beta globin. In Hgb H disease, the amount of alpha globin made by the body is decreased. The amount of alpha globin to be made depends on the inherited instructions located on each person's genes. Genes are found in every cell of the body (except red blood cells). Usually, a child inherits a total of four genes, two from her mother and two from her father. Hgb H disease occurs when the child inherits only one alpha globin gene instead of four.



If you come into contact

with any of the listed medications and/or chemicals, or if you experience high fever and/or any of the symptoms listed below, please contact your physician or health care provider immediately.

or call Children's Hospital Oakland *Thalassemia Nursing*
(510) 428-3347

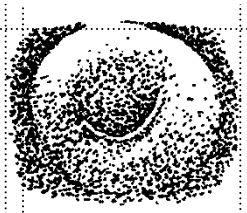
Some common signs of severe anemia include:

- pale or yellowish skin
- yellow eyes
- extreme fatigue
- abdominal/back pain
- dark black stools
- dark orange urine

For further information regarding Hemoglobin H disease, please call or write to:

Children's Hospital Oakland
Northern California Comprehensive Thalassemia Center
Department of Hematology/Oncology
747 52nd Street
Oakland, CA 94609

(510) 428-3347 *Nursing/Medical*
(510) 428-3168 *Genetic Counseling*
(510) 428-3885 ext. 4398 *Outreach*



ALPHA THALASSEMIA

Hemoglobin H Disease

What is Hemoglobin H (Hgb H) disease?

Hemoglobin H disease is an inherited blood disorder that affects a person's ability to produce hemoglobin, causing anemia. Anemia is also known as having a "low blood count."

The disease is a form of alpha thalassemia and is very common in people from China, the Phillippines, Thailand, Vietnam, Cambodia, Laos, and other Southeast Asian countries.

What is hemoglobin?

Hemoglobin is a protein that exists inside the red blood cells, carrying oxygen throughout the body. Hgb H disease causes a decreased production of hemoglobin, resulting in anemia. This anemia can cause people to be more tired than normal. However, most people with Hgb H disease are able to grow normally and live into adulthood.

Why is it important to know about Hgb H disease?

Hgb H disease can cause severe anemia if the person comes in contact with certain medications and chemicals *(see list)*. These medications and chemicals can lead to breakage of the red blood cells, and should be avoided by people with Hgb H disease. Severe anemia can also occur if the patient gets a high fever. Temperature should be closely monitotred during illness and infections should be treated immediately to avoid high fevers. If you come into contact with any of the listed medications and/or chemicals, or if you experience high fever and/or any of the symptoms listed on the back, please contact your physician or health care provider immediately.

There is a more serious type of this disease called Hemoglobin H-Constant Spring (H-CS) disease. Individuals with Hgb H-CS disease have a greater chance of serious complications. These may include having an enlarged spleen (possibly requiring medical treatment) and/or needing occasional or ongoing blood transfusions. Special genetic (DNA) testing is needed to diagnose Hgb H-CS disease.

If you have any questions about medications or foods which cause severe anemia, please call your health care provider or the Northern California Comprehensive Thalassemia Center at(510) 428-3247

MEDICATIONS TO AVOID

for patients with Hgb H Disease:

antimalarials
Primaquine*
Chloroquine*
*reduced doses can be given under surveillance if necessary
Hydroxychloroquine sulfate

tuberculosis drugs
Isoniazid
Rifampin

sulfa drugs
Sulfacetamide (eye drops)
Sulfanilamide
Sulfamethoxazole (Gantanol)
Sulfapyridine
Sulfasalazine (Salicylazosulfapyridine)
Sulfisoxazole (Gantrisin)
Dapsone

analgesics
Aspirin*
*acetaminophen safe as an alternate
Phenacetin*
*moderate doses probably safe
Acetanilide

other antibacterials
Nalidixic acid (NegGram)
Nitrofuratonin (Furadantin)
Furazolidone (Furaxone)
Chloramphenicol
ß-aminosalicylic acid
Ciprofloxacin
Doxycycline

folic acid antagonists
Pyrimethamine

miscellaneous
Vitamin K analogues*
*1mg Menadiol ok parenterally
Phenazopyridine (Pyridium)
Toluidine Blue (a dye)
Methylene Blue (a dye)
Trinitritoluene (TNT)
Quinidine Gluconate
Naphatalene

food
Fava Beans

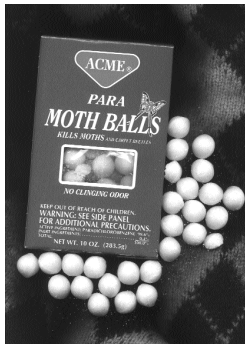
source:
Bull WHO 1989,Beutler 1994

Take this list of medications to any doctor appointments or emergency room visits you may have. It is important to show this list to your health care provider before s/he prescribes any medicine for you.

Compiled by Nicole Heer, MS and Kathy Wong, RN; 1996

Moth Balls and Fava Beans are two other substances which can cause severe anemia and should be avoided.

Moth Balls (Naphatalene):



Accidental swallowing of moth balls or inhalation of its fumes by children with Hgb H disease can be especially harmful, causing severe anemia. Do not keep moth balls in your home. If your child should accidentally swallow a moth ball, call your health care provider immediately.

Fava Beans:

A fava bean is a large bean which can be eaten boiled, sauteed or deep fried. These beans are especially harmful and can cause severe anemia in persons with Hgb H disease. Their particular metabolism causes the fragile wall of the red blood cell to break and lose all of its hemoglobin. Pollen from a fava bean plant should also be avoided. Other types of beans—such as black beans, lentils, peas or string beans—are not harmful.