

Are there special tests for iron overload?

Blood tests for serum iron and either total iron binding capacity (TIBC) or transferrin are good screening tests. The ratio of serum iron or transferrin or TIBC is normally about 0.30 or 30%. Figures above 50% (indicative of iron overload) or below 15% (iron deficiency) need more study. A good additional test is serum ferritin level, which is elevated in patients with hemochromatosis. If these tests are persistently high, a genetic test for the mutations in the HFE gene should be performed. A genetic test is commercially available and costs about \$175. Depending on whether there is evidence of liver damage, a liver biopsy should be done to assess the damage to the liver. Excess iron is also frequently present in patients with alcoholic liver disease or chronic viral hepatitis. A liver biopsy is the only definitive way to determine if patients with these diseases also have iron overload.

How can hemochromatosis be treated?

Phlebotomy treatments (blood letting) are performed where one to two pints of blood (which contain iron in hemoglobin in red blood cells) are removed each week until iron stores go down to a normal level. It may take several months to several years to remove all excess iron. After the iron stores are reduced to normal, maintenance phlebotomy treatments should continue every two to four months for life to prevent re-accumulation of iron.

What is the outlook for patients?

Those who are treated early can look forward to a completely normal life. When the illness has advanced to the stage of cirrhosis, the situation is more serious. Liver cancer can occur in up to 30% of these patients. Damage to the pancreas by the excess iron can result in diabetes mellitus. Damage to other organs may cause arthritis, loss of body hair, heart problems, loss of sex drive (males), and impotence. In the latter stages of the disease, the patient may develop an enlarged liver (hepatomegaly), cirrhosis (scarring of the liver), and an enlargement of the spleen (splenomegaly).

Does having anemia rule out iron overload?

No. There are many forms of anemia, and a person can have both anemia and iron overload.

What effect does alcohol have on hemochromatosis?

Drinking alcoholic beverages should be avoided. Alcohol can accelerate liver damage in those with hemochromatosis.

Is there any relationship between diet and iron overload?

Hemochromatosis is unrelated to diet. It is rare for people to develop iron storage problems after taking large amounts of iron tonics and medications over a long period. However, no one should take iron supplements without a doctor's advice. Anyone with an iron overload problem should avoid tonics and medications containing iron.

Can diet help?

When hemochromatosis is diagnosed, a well-balanced diet, low in iron-rich food, is recommended. Consumption of iron-rich foods such as red meat should be limited to small quantities. Liver, a food high in iron content, should be avoided as it may enhance iron absorption from food. Read food labels carefully, as many foods, such as cereals, are fortified with iron.

People with hemochromatosis are considered to be at high risk of the bacteria *vibrio vulnificus*, found in shellfish, and are therefore advised to avoid eating raw oysters and clams.

Should damage appear in other organs, further dietary recommendations may be indicated:

- Damage to the pancreas will likely require a diabetic diet in which the main focus is avoidance of concentrated sweets. A high-fiber diet may be a benefit to help control blood sugar levels.
- Cardiac problems may require sodium restriction.
- Diabetes mellitus and cardiac problems related to obesity require a weight reduction, low-fat diet.
- Cirrhosis may require restriction of sodium.

Liver transplantation and hemochromatosis

Patients who undergo liver transplantation for hemochromatosis generally have a one-year average survival rate of about 50%, which is 30% lower than those transplanted for other reasons. The lower rate is believed to be caused by infectious or cardiac complications. The survival rate is also lower for patients whose hemochromatosis is not diagnosed prior to liver transplantation for other reasons.

For further information, contact:



American Liver Foundation
75 Maiden Lane, Suite 603
New York, NY 10038

1-800-GO-LIVER (465-4837)

Website: www.liverfoundation.org

E-mail: info@liverfoundation.org

The American Liver Foundation is a nonprofit, national voluntary health organization dedicated to the prevention, treatment, and cure of hepatitis and other liver diseases through research, education, and advocacy.

The information contained in this brochure is provided for information only. This information does not constitute medical advice and it should not be relied upon as such. The American Liver Foundation (ALF) does not engage in the practice of medicine. ALF, under no circumstances, recommends particular treatments for specific individuals, and in all cases recommends that you consult your physician before pursuing any course of treatment.

*The American Liver Foundation gratefully acknowledges the contribution of Bruce R. Bacon, MD, who reviewed and updated the information contained in this brochure.

What is Hemochromatosis?

Hemochromatosis

Hemochromatosis is an inherited condition that causes the body to absorb and store too much iron.

Hemochromatosis is the most common inherited disorder affecting more than one million people in the United States who carry both copies of the abnormal gene. It is estimated that 10% of the population are carriers of one copy of the abnormal hemochromatosis gene. Many cases go undiagnosed because neither patients nor physicians have been alerted to the problem. The most vital factor in making an early diagnosis is enhanced recognition of the disease by doctors and patients. This is particularly important since early diagnosis and prompt treatment can prevent all of the long-term complications of the disease.

Who is most likely to get hemochromatosis?

The hemochromatosis gene, called HFE, was identified in 1996 and is inherited from both parents. It is most often diagnosed in people who exhibit symptoms and are between the ages of 40 and 60 years old. Women who lose iron through menstruation, pregnancy, and breast-feeding often develop symptoms at a later age than men. Anyone who has a blood relative with hemochromatosis should be tested even if there are no symptoms.

What are the symptoms?

Many people have no symptoms, even in advanced cases. Others suffering from hemochromatosis frequently exhibit these symptoms:

- Fatigue
- Weakness
- Abdominal pain
- Pain in the joints
- Slightly elevated liver enzymes
- Bronze or greyish discoloration of the skin
- Impotence