

DIET

It is not possible to eliminate iron overload by a low iron diet because very many foods contain iron. A good balanced diet is recommended. The amount of iron absorbed from most foods is very small compared with the iron removed in a single venesection.

The rate of iron absorption can be slowed by:

- avoiding vitamin supplements or tonics containing iron, and breakfast cereals heavily fortified with iron. Large doses of vitamin C should also be avoided, as it increases the absorption of iron from the diet and makes the process of depositing iron in some organs easier.
- limiting alcohol intake, particularly with meals, as it increases iron absorption and it can also cause liver disease.
- tea and dairy products taken with a meal reduce the amount of iron absorbed.
- cutting down on red meat: iron is much more readily absorbed from meat than vegetables, fruit, cereals and beans.

Disclaimer: This summary information has been approved by our Medical Advisers but is not a substitute for full guidance. Please contact the Society or your Doctor if you require further information or are in any way concerned about your health.

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AIMS OF THE HAEMOCHROMATOSIS SOCIETY

The Society was set up to provide support and information for those affected by GH. Members receive treatment record cards, a handbook, quarterly newsletters and have opportunities to meet other members and specialists locally and at our meetings.

- To **SUPPORT** people with GH by providing them with information about the condition, helping with their problems and encouraging prompt testing of their relatives.
- To promote **AWARENESS** among the health professions, patients and their families, the general public and policy makers so that the condition may be diagnosed and treated in time. There is also a need to overcome the misconceptions that GH is rare, that only middle-aged men are at risk, and that women are seldom affected until their menopause.
- To encourage and support **RESEARCH**, and provide resource material for the allied medical professions.

The Society is a registered charity, and a member of the following organisations, the International Association of Haemochromatosis Societies, the Genetic Interest Group, Contact a Family. It is also affiliated to the British Liver Trust.

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Haemochromatosis

An Iron Overload Disorder

Genetic Haemochromatosis (GH) is an inherited disorder which causes the body to absorb too much iron from the diet. The excess iron gradually accumulates, usually in the liver, pancreas, joints, heart or endocrine glands causing serious tissue damage.

It is now recognised as being one of the most common genetic disorders. Surveys have shown that 1 in 200 people are likely to be at risk of developing iron overload. Diagnosis can be confirmed by a simple blood test.

Simple and effective treatment is available, but if the excess iron is not removed irreversible damage can eventually occur, especially in the liver.

Early diagnosis and treatment preserves normal life expectancy and good quality of life.

SYMPTOMS

Symptoms develop as iron slowly builds up, so are more likely to occur during adult life. Some symptoms can also be caused by other illnesses, but a combination of two or more of the following indicates that GH should be considered.

Arthritis in any joint but particularly in the knuckle and first joint of the first two fingers; this is highly suggestive of GH

Lethargy/feeling tired all the time chronic fatigue, weakness

Abdominal pain in the stomach region, upper right hand side, or diffuse

Mood swings impaired memory, irritability, depression

Reduced sex drive impotence in men, early menopause in women

Skin colour change permanent tan, paleness, or greying

Shortness of breath/heart irregularity

Diabetes (late onset type)

Liver disorders abnormal liver function tests, enlarged liver, cirrhosis

DIAGNOSIS

GH is diagnosed through blood tests which your doctor can order, they are not routine blood tests.

1. Transferrin Saturation (TS) – indicates how much iron is readily available for use in the body. The average is 30% (slightly higher in men than women). If on two occasions this is over 50% in men or 45% in women, GH is very likely. This test is not often requested but it is the most specific and sensitive test for GH.

2. Serum Ferritin (SF) – indicates the amount of iron stored in the body. Levels significantly over 300µg/l [micrograms per litre] in men and post menopausal women and 200µg/l in younger women suggest GH. In the early stages of iron accumulation serum ferritin may be normal.

3. Gene Test – a simple blood test for the HFE gene mutation is positive in over 95% of people with GH. It will identify family members at risk of loading iron even before they have increased body iron stores.

4. Liver Biopsy – this is only usually requested if GH is diagnosed by the gene test and the serum ferritin level is over 1000µg/l, the liver function tests are abnormal, or the gene test is negative. A biopsy shows if the liver is damaged.

If tests indicate GH is likely a referral to a Haematologist (blood specialist) or Gastroenterologist (Digestive system specialist) is usually the next step.

TREATMENT

Treatment is simple and effective, and consists of regular removal of blood. Known as venesection therapy or phlebotomy, the procedure is the same as for blood donors. Each pint of blood removed contains 200mg of iron. The body then uses some of the excess stored iron to make new red blood cells.

Initially venesection will usually be performed around once a week, and continues until the iron level is satisfactory, which may be a year or more. During the course of treatment the size of the remaining iron stores is monitored.

Treatment should usually continue until the SF level reaches 20µg/l, indicating minimal or absent iron stores.

After the stores have been depleted a second phase of treatment is usually required. Iron levels should be monitored regularly, and if necessary occasional venesections continued to keep iron levels down.

The TS should be maintained below 50% and the SF below 50µg/l.

EFFECTIVENESS OF TREATMENT

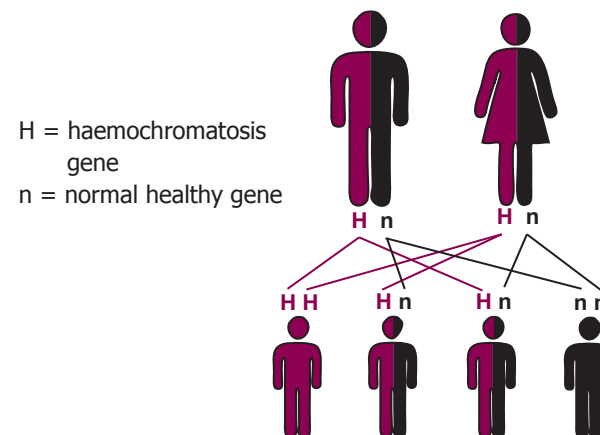
Early diagnosis and treatment is vital. Venesection leads to the reduction in the body's stores of iron to the normal range,

and many symptoms will improve. Providing this begins before cirrhosis has developed life expectancy will be normal. It will not cure some serious conditions caused by iron overload such as diabetes or cirrhosis if they are already present at the time treatment is started.

HAEMOCHROMATOSIS INHERITANCE

To develop GH a defective HFE gene must be inherited from both parents. People who have inherited a defective gene from only one parent are carriers and will not usually be at risk of iron overload. Their children will have a 1 in 4 chance of also being carriers. If both parents are carriers, their children will have a 1 in 2 chance of being carriers and a 1 in 4 chance of being at risk of developing GH. It is estimated that 1 in 8 people in the UK are carriers. For further information please see our website or contact the Society.

How GH inheritance works when both parents are carriers



HAEMOCHROMATOSIS AND FAMILIES

Because GH is inherited it can be passed on from parents to children. When one member of a family is diagnosed close relatives should be informed so that they can get themselves tested. Discuss this with your doctor.