Canadian HEMOCHROMATOSIS SOCIETY
Société canadienne de l'hémochromatose

Hemochromatosis - Treatment

Aggressive De-Ironing

Treatment for hemochromatosis involves management of complications, screening for liver cancer, avoidance of supplemental iron and appropriate vaccinations for hepatitis A and B; however, **an aggressive de-ironing protocol is most important**. **Excess iron is removed by a procedure known as phlebotomy**, which is the drawing off of a unit of blood, using the same technique as a blood donation, but with a much higher frequency.

This treatment is effective because phlebotomies remove red blood cells that contain iron. Each unit of blood contains approximately 225 mg of iron in hemoglobin, the main component of red blood cells. In the process of making new red blood cells, a signal is sent for the stored iron in the tissues and organs to be pulled out and transported to the bone marrow where red blood cells are produced. This repeated procedure gradually depletes the stores of excess iron and eventually the iron levels fall back to normal.

In the initial phase of de-ironing, depending on the amount of stored iron, phlebotomies may need to occur once a week or even twice a week for one year. These **phlebotomies can take place at a doctor's office or a local hospital outpatient clinic**.

Monitoring Progress

Some doctors use hemoglobin exclusively during this phase to monitor progress and establish frequency. It is checked at each treatment and de-ironing is deemed complete when the hemoglobin level falls to 10.5 - 11 g/dL and a mild anemia develops. Patients may become quite fatigued when they are finally iron deficient. In addition to monitoring hemoglobin levels, some doctors monitor ferritin levels periodically during this phase as well and may move to the maintenance phase of treatment when ferritin levels drop below 50 ng/ml.

During the maintenance phase of treatment, the goal is to keep transferrin saturation between 30-40% while maintaining a normal hemoglobin (normal hemoglobin range is 13-18 g/dL for men and 12-16 g/dL for women). **It depends on the individual, but typically this could be achieved with one phlebotomy every 3-4 months. The treatment is ongoing for life**.

Becoming a Regular Blood Donor

If a person with hemochromatosis is otherwise eligible, he / she can **become a regular donor at Canadian Blood Services (CBS)**. Many healthy hemochromatosis patients find the CBS a much more comfortable environment for lifetime maintenance phlebotomy treatment; not only is it therapy, but also it provides much needed blood for other Canadians. Blood donations can be made every 56 days, provided the hemoglobin is normal and the patient is not on insulin.

Phlebotomies can be inconvenient, invasive, and painful for some, and it is a life-long commitment, but they work! **Studies clearly show that both survival rates and quality of life are significantly improved with phlebotomy therapy**, even for patients who have already sustained organ damage. For those diagnosed early, before clinical symptoms manifest, and before there is evidence of liver dysfunction or cirrhosis, longevity is statistically the same as the average person without hemochromatosis, in the absence of other risk factors for liver disease, such as alcoholism or hepatitis.

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Improvement Over Time

Depending on the timing of diagnosis and treatment, many of the disease symptoms will disappear after iron levels are back to normal. Fatigue and depression seem to improve. Some liver function can be improved, but not cirrhosis. As such, those patients require life-long screening for hepatomas. There is a mild improvement in diabetes and heart function. Improvement in the joint symptoms is variable but there seems to be no improvement in some of the hormonal complications, such as impotence. Even if there is no improvement, **preventing progression of the disease process in damaged organs makes phlebotomy treatment worthwhile**.

Diet

Despite that hemochromatosis is caused by inappropriate iron absorption from dietary iron and resulting iron overload, **reducing dietary iron to a below-normal level does not significantly affect treatment** of the disease. However, it is important to modify lifestyle and diet in order to not take in more-than-normal amounts of iron through the diet such as in certain vitamin pills or fortified foods.
See "Treatment of Iron Overload: A Hemochromatosis Primer" in the Fall 2005 issue (www.ironoverload.ca/newsletter/2005_fall.pdf) of Iron Filings, the CHS newsletter, for more on lifestyle and diet issues.

Chelating Agents

For those patients who cannot tolerate phlebotomies, there are chelating agents that will also reduce body iron stores. However, toxicity is a concern, they are not as efficient, and they are inconvenient to administer because they must be injected subcutaneously over several hours. Because of all these factors, they are prescribed only as a last resort. Research with newer versions of chelators and other forms of oral drugs is ongoing. In the future, patients with hemochromatosis may have other options for the type of treatment that is best for them, but for now, phlebotomy is the gold standard.