

For Your Physician

Top five most frequently asked questions about Hemochromatosis and Iron Disorders Institute (IDI) recommendations

1. **Is the compound heterozygote at risk of iron loading?** Not enough is known about the loading patterns and penetrance of heterozygotes to discount the potential risk of morbidity/mortality in this population. Until more evidence is available, **Iron Disorders Institute (IDI) recognizes the compound heterozygote (C282Y/H63D) in the same risk group** for iron loading as the C282Y homozygote, although iron loading may be less severe in the compound het population.
2. **Do I have to become anemic to be de-ironed?** Forced-sustained anemia is outdated and harmful. IDI recommends **a pre-pheraphotomy hemoglobin of 12.5g/dL (Approx HCT 37.5%) to avoid overbleeding**. There are exceptions, which are emphasized on the enclosed Physician's Hemochromatosis Reference Chart. IDI's recommendation is upheld by the National Institutes of Health Hemochromatosis Protocol, Bethesda, MD.
3. **Do I need a liver biopsy? Liver biopsy is no longer used to diagnose hereditary hemochromatosis;** but is recommended for HHC patients with serum ferritin $\geq 1,000\text{ng/mL}$ at the time of diagnosis. Serum ferritin is an important part of the treatment and iron reduction management, falling $\sim 30\text{ng/dL}$ per 500cc of blood removed. Serum ferritin is also an acute-phase reactant and can be elevated in conditions of inflammation or ingestion of certain drugs such as nicotine, alcohol or estrogens. This should be taken into account for individuals whose ferritin falls in large increments.
4. **What about my children?** In families where HHC is diagnosed, for children under the age of 18 but no younger than age 3, IDI recommends **a bench-mark iron panel** (fasting serum iron, TIBC or UIBC and serum ferritin) **at the first opportunity to draw blood e.g., back to school physicals**.
5. **What about diet and supplements? Reduction of consumption of alcohol, red meat, fats and sugars are recommended. Raw shellfish should be avoided.** Supplemental vitamin C should not exceed 200mgs per dose and calcium supplements which are known to reduce heme and non-heme iron absorption but should not be used in place of TP. **A daily multiple vitamin without iron and fluids** are recommended for patients undergoing TP. Excessive supplementation is not recommended by IDI but vitamin B complex and silymarin are among the supplements considered beneficial for HHC patients. **Fresh fruits and vegetables including spinach are encouraged regardless of vitamin C content.**