Phlebotomy Guidelines for Patients with Hereditary Hemochromatosis

**Hgb** | **SF** | **TS%** | **Action Employed** | **MCV** | **Comments**
--- | --- | --- | --- | --- | ---
Normal | Elevated** | Elevated greater than 45% | Aggressive | One (500cc) or two units per week (depending upon initial SF and alcohol consumption) until SF is lowered to ~750 ng/mL. Consider DRCA (double red cell apheresis). | Normal | With few exceptions, pre-treatment Hgb should be 12.5g/dL or greater. SF and TS% should be checked once a month during iron reduction phase. CBC may also be done at this time to determine MCV, etc.

**Important:** Serum ferritin (SF)>1,000ng/mL is CRITICAL and requires aggressive treatment. The risk of cirrhosis is <1% in patients whose SF has not been elevated above 1,000 ng/mL.

**Normal** | Elevated** above normal up to 1,000 ng/mL | Elevated greater than 45% | Aggressive to Moderate | One unit (500cc) per week depending upon patient, may need to adjust to one unit every other week. | Normal | MCV will drop by 3% of baseline without causing anemia, when iron reduction reaches ideal SF (see chart below) and TS% levels.

**Normal** | High Normal** | Elevated greater than 45% | Standard Therapeutic Phlebotomy | One unit (500cc) monthly. | Normal | Well tolerated by many patients; availability varies for different advantages; smaller needle and fewer extractions.

**Normal** | Normal 25-35% | Routine Blood Donation | One unit (500cc) every 8 weeks, to maintain SF 50-150 ng/mL with TS% <40% | Normal | Some patients may need fluid replacement.

**Normal** | Elevated** | Normal | Rule out NASH, diabetes/mTOR overload syndrome (DIOCS) chronic liver disease (alcohol, hepatitis) or hyperferritinemia cataract syndrome (HFC). NASH diagnosis includes hypertransaminasemia + hepatic index >1.9. HFC: diagnosis: ophthalmologist confirms early onset cataracts. HFC is not a condition of iron overload. | Normal | Diet during iron reduction should include restriction of red meat; no raw shellfish or alcohol.

**Normal** | Elevated** | Elevated | Common phenomenon for patients with classic hemochromatosis, possibly caused by abnormal shuttling of iron into plasma due to genetic makeup of the patient. Discontinue phlebotomy until SF rises to ideal range. Read about Iron Avidity: www.ironorders.org or www.hemochromatosis.org | Normal | On ideal ranges are achieved, providing limitations of disease consequences (diabetes mellitus, renal disease, etc.) the diet may resume without restriction.

**Normal** | Elevated** | Elevated or normal | Rule out anemia of chronic inflammation | Elevated | Single Topical Handouts for your patients: Hemochromatosis Diet Hemochromatosis Genetics Iron Avidity DIOS How Much Alcohol is Safe? | Elevated | See maintenance below

**Sample Phlebotomy Order**

"Phlebotomize 500 cc once a week** if Hgb>12.5g/dL** (Approximate hematocrit of 38%)" period of time should reflect frequency desired

Note: For patients whose initial ferritin is greater than 1,000ng/mL, SF should be evaluated every 4-6 weeks until lowered to ~200 ng/mL. Otherwise, SF and TS% can be checked every two bleeds until SF reaches 75ng/mL.

**Key References:**

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The therapy for the majority of hemochromatosis patients is blood donation or phlebotomy. Most patients are candidates for standard phlebotomy. With few exceptions, patients should have a pretreatment hemoglobin of 12.5 g/dL. Quantities removed by phlebotomy may vary from minimal extraction of 250cc up to large volume extraction of 600cc or more with double red cell apheresis (DRCA). Extraction continues until ferritin reaches 25g/mL on one occasion but hemoglobin does not drop below normal range for age, weight or gender.

Alcohol consumption or concurrent disease: will influence a patient’s unloading pattern and phlebotomy frequency may need to be adjusted. Patients whose initial serum ferritin is above 1,000ng/mL should be advised to abstain from alcohol. A patient’s liver health and risk of alcohol consumption should be determined by the physician.

Maintenance: Once iron reduction is achieved (ideal SF and TS% ranges are reached) the patient may require as few as 3 or 4 phlebotomies a year. Patients can be advised of diet and techniques to help reduce the amount of iron absorbed, which may lessen the frequency of phlebotomy during maintenance.

Minimal extraction: used for youths, persons who are frail, small in stature or weight, or who have coexistent illness such as heart problems Blood is extracted from vein in the arm using a 20-22 gauge butterfly needle with vacuum bottle. Duration is about 15-20 minutes. The amount extracted is about 250-300 cc (about 125 mg of iron). The frequency can adjusted depending on patient tolerance. Other patients may benefit from this method of blood removal are those with small, inaccessible, scarred or rolling veins or those who cannot tolerate a standard phlebotomy.

**Important Ferritin Reference Ranges**

<table>
<thead>
<tr>
<th><strong>Ferritin</strong></th>
<th><strong>Adult Males</strong></th>
<th><strong>Adult Females</strong></th>
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</thead>
<tbody>
<tr>
<td>Ideal Range</td>
<td>50-150 ng/mL</td>
<td>50-150 ng/mL</td>
</tr>
<tr>
<td>Induction Phase***</td>
<td>50-75 ng/mL</td>
<td>50-75 ng/mL</td>
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</tbody>
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Serum ferritin decreases ~30ng/mL per 500cc phlebotomy (Harrison, et al)

**Adolescents, Juveniles, Infants & Newborns of normal height and weight for their age and gender**

| **Male ages 10-19** | 23-70 ng/mL | **Female ages 10-19** | 6-40 ng/mL |
| **Children ages 6-9** | 10-55 ng/mL | **Children ages 1-5** | 6-24 ng/mL |


**Note:** These guidelines are intended to reduce the risk of under-bleeding or over-bleeding a patient. Each patient is unique. Clinicians may need to adjust phlebotomies accordingly.

**Treatment Centers:** Iron Disorders Institute maintains two websites that provide information about the National Network of HHC Donor Centers. Many of these centers have a variance from the FDA to use HHC blood for transfusion, and do not charge the patient for phlebotomy, regardless of the frequency. A physician’s order is necessary for the patient to participate. Any HHC patient who lives near Bethesda, MD may wish to contact The Warren Magnuson Clinical Center, Hemochromatosis Protocol Coordinator, Yu Ying Yau, RN, at 301-496-1431 or Y.Yau@nih.gov www.ironorders.org or www.hemochromatosis.org