



## Appendix F: Management and surveillance of *HFE*-HH

### ► Therapeutic Phlebotomy

- Phlebotomy is the treatment of choice for HH and for other primary iron overload disorders. Possible exceptions include those who are already anemic or have limited life expectancy due to other diseases or those with congestive heart failure with hemodynamic instability.
- Volume and frequency of phlebotomy need to be individualized according to the patient's age and clinical and biochemical presentation. Some patients will tolerate weekly phlebotomy but most will not tolerate phlebotomy more frequently than every 2 – 4 weeks and smaller volumes may be indicated in women. Guidelines recommend that for severely iron overloaded patients, weekly phlebotomy of 500 ml of whole blood should be continued until serum ferritin is 50-100 µg/L, within the patient's tolerance. Consider venous access, frequency of phlebotomy, and symptoms of iron deficiency). Patients with massive iron overload may require in excess of 100 phlebotomies.
- Serum ferritin and hemoglobin should be monitored regularly (e.g., every 4th phlebotomy) to assess response to therapy. It is unusual for iron overloaded patients to develop anemia early in the course of phlebotomy therapy. If this occurs, reduce the frequency of phlebotomy.
- Once patients have been successfully depleted of excess iron stores (ferritin <50 µg/L), a program of monitoring and maintenance should be established. The need for maintenance phlebotomy is quite variable; some patients require phlebotomy every 2 - 4 months to maintain a low-normal ferritin, and some may not re-accumulate for many years.
- At minimum, the CBC, ferritin and TSAT should be checked every 12 months, and phlebotomy should be re-initiated if the ferritin or TSAT is rising toward the upper limit of normal.
- People with hemochromatosis can donate blood, provided they meet all other Canadian Blood Services donor eligibility criteria. See details from Canadian Blood Services at: [blood.ca/en/blood/am-i-eligible/abcs-eligibility/hemochromatosis](http://blood.ca/en/blood/am-i-eligible/abcs-eligibility/hemochromatosis)

### ► Chelation

- Patients with *HFE*-HH respond to phlebotomy and do not require chelating agents such as deferasirox and desferrioxamine.

### ► Organ Damage

- In patients with organ damage, such as cirrhosis, congestive heart failure, hypogonadism, referral to the relevant specialist is recommended.
- End organ damage should be reassessed periodically. Reversal of organ impairment is sometimes seen once iron stores have been depleted. For example, if liver enzymes have been abnormal, they often improve with phlebotomy. There may also be improvement in iron-induced cardiac dysfunction. Diabetic patients often note improvement in blood sugars with less dependency on insulin or oral hypoglycemic agents. Conditions that often do not improve with phlebotomy include arthropathy, cirrhosis and testicular atrophy.

### ► Dietary Restriction

- Strict avoidance of dietary iron is not necessary, but iron and vitamin C supplements should be avoided.<sup>17</sup> Patients can access the support of a dietitian through HealthLink BC.