ABSTRACT

Patients are often referred to the Blood Bank for therapeutic phlebotomy treatment of systemic iron-overload and secondary polycythemia. This report summarizes the 20+ year experience of a laboratory-based Hemochromatosis Clinic functioning as a service of the hospital's Blood Bank.

Patients are referred to the "Clinic" by primary care physicians or gastroenterologists, with an order for therapeutic phlebotomy or "Hemochromatosis protocol" and are interviewed for pertinent history and patient questions by the Clinical Pathologist as Clinic Director. Confirmatory and baseline lab tests are performed and a treatment schedule planned and discussed with the patient and often with the spouse and family.

A Consult Letter to the referring physician outlines the treatment plan, asking for comments and suggestions. A Flow Chart of treatments and lab monitoring data is maintained and periodically sent to referring physicians with updated treatment plans. Overall care of the patient remains the responsibility of referring physicians.

Diagnostic confirmation and treatment planning follows published guidelines, with evidence of systemic iron excess considered if the serum ferritin level above the age-gender reference range, AND the transferrin saturation is greater than 45%, results confirmed by repeat testing, preferably on a fasting sample. Other baseline labs include a CBC, transaminase levels and a CRP level. HFE genetic testing is obtained for patient and family counseling.

Treatment schedules depend on the severity of iron excess and general patient condition. A more frequent Induction Phase is necessary to treat patients with a significant iron overload, tapering the schedule to a patient-variable Maintenance Phase as ferritin levels decrease to a target range.

Patients are advised of a life long need for awareness of their "Iron Status", with lab monitoring and treatments as necessary.

Our Clinic averages 100 patients in its "Hemochromatosis Registry". Patient response has been very positive—related to a dedicated team of therapists and ready access to physician discussion of the nature of their ailment and answers to questions as they arise.

Medical Staff response has been very positive as they are relieved from responsibility for "day-to-day" patient monitoring and scheduling.

Clinical Pathologists have a unique opportunity & responsibility for direct patient care with an ongoing doctor-patient relationship.

CLINICAL PATHOLOGIST'S ROLE:

As a clinician specialized in diagnostic confirmation and treatment of referred patients with presumptive systemic iron overload. The referring physician retains overall patient care responsibility.

HEMOCHROMATOSIS CLINIC: The Patient-care Process

1) Primary care referral of patient to Hemochromatosis Clinic w/ presumptive diagnosis of Iron Overload.

2) Confirmation of Iron Overload Diagnosis: Baseline lab testing. [One-third of patients don't confirm!]

3) Patient interview: Personal & family history; Discussion of iron overload disorders—diagnosis & Treatment Plan; Patient questions. (Spouse participation encouraged.)

4) Consult Letter to referring physician w/ findings & proposed treatment plan, w/ request for "Comments and suggestions".

5) Induction-phase of treatment, w/ patient interviews p.r.n.; Chart review & re-evaluation each visit.

6) Treatment Flow-chart & updated Consult Letters to referring physician as appropriate.

7) Maintenance-phase of treatment, w/ patient counseling as treatment intervals increase.

ADVANTAGES of the Clinic Protocol:

Improved patient care with:

Unified treatment plan and dedicated patient-care team; Ongoing physician-patient interface—Discussions and questions; Continuous over-view from patient referral to long-term maintenance; Elimination of unnecessary treatments—unconfirmed referrals; Correction of mistaken orders—insuring necessary treatments; Management of H&H variations w/ flexible scheduling.

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SELECTED CASE SUMMARIES

69M: Classic case: Abnormal LFT's w/ cholecyctectomy; Ferritin 2834; 100% sat; C282Y/C282Y; 29 phlebotomies in 12 mos; Ferritin 135; AST/ALT normal. Phasing to maintenance schedule.

52M: NHL treated to remission—no iron studies done; Notified by brother to get tested for hemochromatosis; Ferritin 1162, 83% sat; C282Y/C282Y; 14 phlebotomies in 12 mos; Ferritin <100; In maintenance phase.

75F: Liver biopsy w/ advanced cirrhosis & 4+ iron; Negative for 3 HFE mutations; African American; Life-long history of excess iron ingestion; Ferritin 8404 w/ 91% sat;152 short-draw phlebotomies in 8 yrs w/ Ferritin stable for 2 years @ 7-20, 25% sat; 70M: Previously treated C282Y/C282Y w/ severe reaction to 500cc draws; Ferritin 1010, 56% sat; Hgb 10.5 w/ B-thalassemia minor; 64 short-draw phlebotomies in 16 months to Ferritin <100; Ferritin stable <200 for ten years; now 83 w/ annual lab monitors.

70F: Transfusion hemosiderosis 12 yr after stem-cell (x2) transplants for AML & "dozens" of transfusions; Neg for 3 HFE mutations; Ferritin 2151, 41% sat; Ferritin 545 @ 16% sat after 11 phlebotomies; CRP 6.7 w/ bilateral hip replacements scheduled. [Residual reactive ferritin elevation related to severe arthritis.]

52F: Cutaneous porphyria. Biopsy proven w/ painful skin lesions; Ferritin 1058, 60% sat; 8 short-draw phlebotomies (petit lady) w/ Ferritin 70, 12% sat and resolution of skin lesions.

REFERENCES:

