Clinical Guidelines

Nurse led Haemochromatosis Service

St. Joseph's Hospital/

Beaumont Hepatology Unit

F Colclough J Ryan 05/06/19

1.0 Title 2.0 Clinical Guidelines for Haemochromatosis	**Date Originally Approved:** **Revised Document Approved:** **Next Revision Due:**

2.0 Scope:

The Haemochromatosis service will ensure all patients with a diagnosis of Hereditary Haemochromatosis and iron overload will access safe and effective treatment in a timely manner with a multidisciplinary approach.

Treatment (Venesection) is indicated:

- C282Y homozygotes: when serum ferritin level is >300 µg/L in men and >200 µg/L in women
- Compound C282Y/H63D heterozygotes: when serum ferritin level is >300 µg/L in men and >200 µg/L in women provided the transferrin saturation (Tsat) is elevated (>45% in men and women)
- H63D Homozygotes: if ferritin >500 and discussed with Lead consultant.

The service is provided daily Monday to Friday, 08:00-13:30.

Location of service: First Floor, St. Joseph's Hospital, Raheny.

The consultants referring their patient to the Heamochromatosis service will be responsible for communicating the clinical guidelines to their teams. **A blue referral card is present in outpatients which must be completed in full, along with the patient's contact number** to facilitate prompt access to treatment.

3.0 Purpose:

The purpose of this document is to guide staff in the management of patients who have a diagnosis of Hereditary Haemochromatosis attending the service at St. Joseph's Hospital, Raheny.

4.0 Definitions:

Haemochromatosis is an autosomal recessive hereditary condition caused by mutations in the HFE gene (mainly C282Y and H63D), whereby excessive iron is absorbed from the diet and deposited in various organs, mainly the liver, pancreas, heart and joints resulting in organ damage and impaired function.

Actions & Responsibilities

1. Governance	Responsibilities:
1.1 Hepatology team Haemachromatosis patient will be admitted under the Hepatology team. The Hepatology team will ensure that all members of the team know the referral pathway. The Hospital Manager knows and is aware of the referral and care pathways.	Dr John Ryan Dr Karen Boland Dr Danny Cheriyan Prof Gavin Harewood Prof Frank Murray Dr Aoibhlinn O'Toole Prof Stephen Patchett Dr Conor O'Brien Ms. Mary Keogh
1.2 Emergency Medical Cover Emergency medical cover will be provided by the medical team in St. Joseph's Hospital. See Standardised approaches to Communicating an emergency for St. Joseph's Hospital Campus.	Prof Ciaran Donegan Dr Alan Moore Dr Alan Martin
1.3 Nurse led service The Haemochromatosis clinic is a nurse led service, with support from Dr Ryan Consultant Hepatologist, Beaumont Hospital. The CNM 1 has overall autonomy and accountability with regards to the caseload and ensuring safe patient care throughout the care pathway. The CNM1 meets with Dr Ryan twice per month to discuss individual cases and any service concerns.	CNM1 Dr John Ryan
1.4 Clerical Support Patients are admitted and discharged through admissions in St. Joseph's Hospital. Clerical support is provided for ordering charts, filing, and coding.	Administrative

2. Clinical guidelines	Responsibilities:
2.1 Management of hereditary Haemachromatosis **Indication for Venesection** On the first visit the CNM 1 will have a patient education session and written information with regards to the condition and the patient will receive a venesection record booklet.	CNM1
2.2 Clinical parameters- treatment initiation Serum ferritin level >300µg/L (males), >200µg/L (females) C282Y homozygotes, and in compound C282Y/H63D heterozygotes with TSat. >45% H63D Homozygote – only if ferritin > 500µg/L and discussed with lead consultant.	
2.3 Hepatology review/Fibroscan request C282Y homozygotes with clinical evidence of liver disease • Persistently abnormal ALT • Initial serum ferritin >1000µg/L • Age: >40yr plus other risk factors for liver disease such as alcohol, Hep B & C, or steatohepatitis Compound or C282Y heterozygote with elevated TS, abnormal LFT's or clinical evidence of liver disease.	
2.4 Therapeutic Venesection Removal of 450ml blood (250g iron), = Serum ferritin drop of 35-50µg/L Aims: 1. Serum ferritin 50-100µg/L (males and females); maintenance phase 50-150µg/L (males and females) 2. Avoid venesection in patients with a transferrin saturation <45% 3. Avoid anaemia, maintain Hb 11.5 -16.0g/dL; ensure packed cell volume does not fall >10% of original value. Neutrophils – low – needs to be highlighted to admitting consultant.	
2.5 Frequency of treatment (venesection) **Reduction Phase I** Initially 1-2 weekly as tolerated by patient	
2.6 Reduction Phase 2 Ferritin <300 µg/L (males) and <200 µg/L (females): can decrease frequency of treatment to 3-6 weekly Ferritin <50-100µg/L requires repeat bloods every 3-4 months	
2.7 Maintenance phase Ferritin >100µg/L (males and females) will attend for maintenance treatment as per clinical guidelines.	

3. Routine Monitoring of Heamochromatosis	Responsibilities:
3.1 Clinical parameters 3.1.1 Check Hb with each venesection, maintain Hb levels at 11.5 g /dL – 16 g /dL 3.1.2 Check serum ferritin: • every 4th venesection • 3-6 monthly during the maintenance phase 3.1.3 Aim: To maintain ferritin • <100 µg/L in reduction phase • <150µg/L in maintenance phase 3.1.4 Assesment for end organ complications: • Rheumatology assessment for joint disease (if symptomatic arthralgia) • Dexa bone scan for osteopenia/osteoporosis 3.1.5 All patients with a ferritin >1000 µg/L should have • Hepatology assessment • HbA1c • ECG • TFTs • Consider investigations for hypogonadism • Consider echocardiogram 3.1.6 Diagnosis of **liver cirrhosis** requires the following investigations • Alpha-fetoprotein and liver ultrasound 6 monthly • Screening OGD for oesophageal varices every 2-3 years • Vaccinate for Hep A & Hep B • Annual influenza vaccine • Pneumococal vaccine every 5-10 years 3.1.7 Lifestyle measures • All patients will be encouraged to restrict alcohol consumption to within recommended limits • Patients with significant liver disease should be advised to be completely abstinent from alcohol • All patients should be encouraged to adopt regular exercise and a well-balanced healthy diet	CNM 1

Monitoring & Evaluation:	
Related Documents: **AASLD Practice Guidelines 2011** **Clinical Practice Guidelines: Management of patients with HFE-related heamochromatosis (2010)** **HSE Haemochromatosis patient information booklet** **Infection control guidelines** **Venepuncture guidelines** **Communication protocol in the case of an emergency** **Guidelines on a Cardiac arrest**	
Approved By:	**Date Approved:** **Next Review Date:**