

BRISTOL HAEMATOLOGY UNIT

STANDARD OPERATING PROCEDURE

TITLE: HAEMACHROMATOSIS VENESECTION

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1. INDICATIONS FOR PRACTICE

- 1.1 The ferritin level from the previous admission and the haemoglobin are greater than indicated on the patient's protocol.

2. AUTHORISED PERSONNEL/TRAINING REQUIRED

- 2.1 Any nurse who has attended the Venepuncture Study Day and has been assessed as competent according to AHU guidelines.

3. PROCEDURE

3.1 EQUIPMENT

The following equipment is required to perform the procedure:

- Blood Pressure recording machine
- Blood bottles and forms
- Venesection blood collection bag
- Spring balance
- Disposable gloves
- Tourniquet
- Alcohol wipe
- Tape
- Cotton wool/gauze

3.2 PREPARATION

1. Record the patient's pre venesection blood pressure and pulse to ensure this is within safe limits. If systolic is less than 100 or greater than 170 or the diastolic is more than 100 consult the doctor.

ISSUED BY:	NAME	DATE
PROGRAMME QUALITY MANAGEMENT ADMINISTRATOR		22 nd March 2016

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2. Check medical notes/patient progress form to monitor the stability of the patients haemoglobin and to see if a full blood count is required prior to venesection, if so perform accordingly. Obtain result from Sysmex machine. If result is consistently within the criteria stipulated on their progress sheet then a full blood count and ferritin, if required, can be performed during the venesection process and reviewed retrospectively. Record the results on the progress sheet in the patient's medical notes.
3. Proceed to venesection if the ferritin level and red cell indices fall within the criteria indicated on the patient's protocol. If in any doubt discuss with the Doctor.
4. If venesection is not required discuss date of next appointment with patient and doctor, if unsure, and book the patient onto the electronic diary, Mosaiq. Complete paperwork in medical notes and complete coding form.

3.3 PROCEDURE

1. Check the patient's protocol to see if the patient requires an isovolaemic venesection and if so a cannula should be sited into a vein on the opposite arm to the venesection site. Infuse 500mls normal saline while the venesection is in progress as a rapid infusion. The fluid must be prescribed in accordance with trust policy.
2. Explain the procedure to the patient before commencing to ensure the patient is aware of forthcoming actions.
3. Lie the patient down in a semi-prone position supported with pillows on a bed/reclining chair. The reclining position is in case the patient feels unwell and becomes hypotensive during the procedure due to the blood being venesected.
4. Wash hands and put on disposable gloves as per trust policy.
5. Prepare for the venesection by hanging the blood collection bag on the suspended spring balance.
6. Identify a suitable vein for venepuncture, preferably the anti-cubital fossa as this is easily accessible and can be kept straight and still during the procedure.
7. Use a tourniquet but loosen the pressure once the venesection needle is in position. Use the tourniquet to control flow / pressure.
8. Cannulate the vein with the needle attached to the blood collection pack.
9. Check the blood flow to ensure the needle is correctly positioned and that the tubing is patent and blood draining into the bag.
10. When satisfied with the above, secure the needle and tubing to the patients arm with tape, to prevent the needle from falling out.
11. Check the spring balance is weighing the blood in the blood collection bag. Once the required volume of blood has been venesected (as per patient's protocol) and any blood samples that are required have been taken from the vacutainer port of the venesection tubing, release the tourniquet and remove from the patients arm.
12. Remove the needle from the vein, covering the puncture site with cotton wool or gauze and ask the patient to maintain pressure on the site to ensure the bleeding stops. Cover with tape or a plaster.

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13. Pull the blue plastic needle protector over the needle until it clicks in to place, insert the blue needle protector into the vacutainer port to create a closed system. Drain the blood from the tubing into the blood collection bag and put on the clamp.
14. Place the blood bag in the blood bag waste bin.
15. The patient should remain in the reclining position for at least 10 minutes and be offered a drink and biscuits to replace the lost fluid.
16. Check and record the patient's blood pressure post procedure. If systolic is less than 100 or not within patient's normal limits discuss with the day unit doctor. Ensure the patient feels well before they stand to prevent fainting.
17. Record action taken on the patient progress form and in medical notes.
18. Make the next appointment. If unsure when this should be, discuss with Doctor.
19. Complete coding form.
20. Discharge the patient.

4. FURTHER INFORMATION/EXCEPTIONS

Appendix 1, Diagnosis and Management of genetic haemochromatosis

Appendix 2, Haemochromatosis Venesection Management Plan.

DIAGNOSIS AND MANAGEMENT OF GENETIC HAEMOCHROMATOSIS

INTRODUCTION

The defects in the HFE gene, which underlie haemochromatosis, are common within the UK population, but not all individuals carrying the gene defect will clinically manifest the disorder. In those who do manifest the disorder, however, prompt recognition and treatment can help prevent the complications of iron overload. It is, therefore, important that both patients with a high ferritin, who may have haemochromatosis, and family members of known cases, are investigated and managed by the appropriate specialist team.

Investigation

Establish diagnosis

- Fe overload: fasting Transferrin Satⁿ >45% (ideally twice).
- If Ferritin raised with normal Tfn Sat, then consider inflammation, DM, EtOH, liver dis, metabolic syndrome. Check BP, BMI, CRP, γ GT, Glu, triglycerides, cholesterol, ferroportin disease
- HFE gene analysis: usually C282Y homozygote, though rarely due to compound heterozygosity for C282Y/H63D (or C282Y/S65C)
- In iron-loaded patients without C282Y/H63D, consider Fe-loading anaemias (check FBC, retics, film, Hb electrophoresis, ?marrow), or other rare genetic disorders.
- In rare cases without detectable HFE gene mutations, a liver biopsy may be required to establish clinically significant tissue iron overload
- If a genetic cause is thought likely but standard HFE mutations are absent, it may be possible to test for a batch of rarer mutations in other Fe metabolism genes (done by Patricia Bignell in Oxford).
- Complete 'Haemochromatosis Presentation' form (Appendix 1, page 4)

Assess tissue damage

- FBC, Ferritin, Transferrin Saturation
- Liver function tests, including baseline α -Fetoprotein
- Endocrine tests: Glucose, HbA1c, TFTs, Testosterone (\uparrow). Dexascan.
- **Referral for Liver biopsy (or Fibroscan):** if Ferritin >1000 μ g/L, or hepatomegaly, or raised ALT, or diagnosis made > 40 yrs.
- Liver ultrasound if LFTs abnormal
- Joint X-rays of any painful joints
- ECG; consider echocardiogram if any cardiac symptoms/signs

Genetic/Family studies

- Offer Ferritin, Tfn Sat & HFE gene studies to all siblings; document in notes and a clinic letter
- If > 1 child, consider HFE testing other parent before suggesting testing child
- Children should not generally be screened before age 18 yrs (i.e. to be done with their own consent)

Management

'Enhanced liver follow-up' for those with cirrhosis or advanced fibrosis at presentation

- α -Fetoprotein and liver ultrasound, every 6/12 (screening for hepatocellular cancer)
- These patients should be in liver clinic follow-up to arrange these tests

Initial Management of Fe overload (nurse-led service) (Appendix 2, page 9-10)

- Venesect 500ml weekly if tolerated (↓ in elderly, CVS disease or treatment)
- Contact Dr for advice if patient symptomatic post-venesection (may need IV fluid replacement or omission of antihypertensives)
- Monitor FBC with each venesection: reduce frequency of venesection (e.g. to fortnightly) if Hb becomes < 110g/L
- Measure Ferritin mthly; ↓ venesection frequency to monthly when Ferritin <150 μ g/L
- When Ferritin < 70 μ g/L, measure Transferrin Saturation with each venesection.
- When Ferritin <20 μ g/L and Tfn Sat < 20%, start maintenance programme
- Record results on progress chart at each visit
- Patient should be seen in consultant haematology clinic at least every 6 months during this phase, and more frequently if any problems.

Maintenance of normal Fe status (nurse-led service) (Appendix 3, page 11-12)

- Start when patient is confirmed to have low/normal Fe status (Ferritin <20 μ g/L and Transferrin Saturation <20%).
- Dr to estimate venesection frequency that will keep Ferritin 50-100 μ g/L.
- Venesect every 2-4 months, checking FBC, LFTs & Ferritin at each visit.
- If Ferritin rises to > 150 μ g/L, d/w Dr - ↑ frequency of venesection (say to every 2/12).
- If Ferritin falls to <20 μ g/L or Hb falls below normal for pt, d/w Dr - ↓ frequency venesection.
- Patients should be seen annually in Haem OPD; or more frequently if special problems. Note attendance on the follow-up proforma, and check this is at least annual
- If patient on 'enhanced liver follow-up', ensure from time to time that the patient is attending liver clinic and having α FP and ultrasound x2/year
- Discuss with Dr at any time if any queries about management.

References
EASL Clinical Practice Guidelines for HFE Hemochromatosis. J Hepato 2010 doi:10.1016/j.jhep.2010.03.001
http://www.uptodate.com/contents/treatment-of-hereditary-hemochromatosis?source=search_result&selectedTitle=4%7E73
Diagnosis and Management of Hemochromatosis: 2011 Practice Guideline by the American Association for the Study of Liver Diseases. HEPATOLOGY, Vol. 54, No. 1, 2011

1. APPENDICES

Appendix 1 Proforma for use with newly presenting Haemochromatosis patient (page 4)

Attach Banda label

Haemochromatosis Presentation

To be completed prior to referral to Day Unit, and then kept on Day Unit. It should accompany patient at each clinic visit.

Date of first Haematology appointment: _____

Referred from: Gastro / GP / Other : _____

Primary presenting Problem: _____

Problems noted at presentation

Weakness/Tiredness Y / N / N/K
Arthralgia/Arthritis Y / N / N/K
Abdominal pain Y / N / N/K
Impotence (men) Y / N / N/K
Loss of libido Y / N / N/K

Pigmentation Y / N / N/K
Diabetes Y / N / N/K
Cirrhosis Y / N / N/K
Heart Disease Y / N / N/K
Thyroid Y / N / N/K

Alcohol consumption: Current: nil / social / excessive: _____ units/wk
(‘Social’ is <14u in ♀, <21u in ♂) Past: nil / social / excessive: _____ units/wk

Other relevant PMH: _____

Last results prior to venesection: Ferritin _____ µg/L Transferrin sat _____ %
ALT _____ IU/L

Genetic Diagnosis: _____ MCV _____ fL Hb _____ g/l

Haematology/Genetic Management

Health/diet advice given: Y / N

(Normal healthy diet, avoid supplements containing Fe or vitamin C, avoid uncooked seafood when Fe-loaded; consider avoiding Fe-supplemented cereals)

Alcohol advice given: Y / N

(Abstain if LFTs abn; otherwise <14 units in ♀, <21 units in ♂)

Patient information source recommended?: Y / N

<http://www.britishlivertrust.org.uk/liver-information/liver-conditions/haemochromatosis/>

Instruct patient to attend Day Unit for venesection having had normal food & drink that day. For first few times patient should not drive home and should be accompanied. Instruct patient to omit antihypertensives if they have been symptomatic after previous venesections

Genetic screening initiated: Y / N

(Siblings should be offered Ferritin/Tfn saturation & HFE gene analysis. Children are not usually tested – test other parent first if >1 child. Children should not be tested until age 18 anyway). List people offered screening in notes and clinic letter.

Hepatology Management

Refer for consideration of liver biopsy or Fibroscan if any of these apply (tick):

- Ferritin >1000µg/L _____
- Hepatomegaly _____
- Raised ALT _____
- Diagnosis > age 40yrs _____

Date referred to Hepatology: _____

Hepatologist seen: _____

Liver test: Biopsy / Fibroscan / Nil

Advanced fibrosis or cirrhosis? Y / N

(If these are present, patient should remain in liver clinic and undergo αFP & ultrasound 6-mthly). These pts have ‘**enhanced liver follow-up**’.

Other management (date, consultant)

Endocrine referral: _____
(e.g. DM, low testosterone, HypoT4)

Other referral: _____
(e.g. Cardiology, Rheumatology)

Haemochromatosis Venesection Management Plan

Genetic Diagnosis

Presenting Problem

Enhanced liver follow-up Y / N

Induction Phase: Venesect every **weeks**. Aim for ferritin level of **< 20ug/L** for induction phase

Signature of Dr/Nurse Specialist _____ Print Name _____ Date _____

Induction Phase completed: Signature of Dr/Nurse Specialist	Print Name	Date
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Maintenance Phase: Venesect every 4-6 weeks. Aim for ferritin level of 50-100ug/L during maintenance phase

Signature of Dr/Nurse Specialist **Print Name** **Date**

Special requirements e.g IV fluid replacement

Signature of Dr/Nurse Specialist **Print Name** **Date**

Venesect 450- 500 mls each attendance

Monitoring guidance for nurses:

- Repeat ferritin every 4 weeks during induction phase and each visit whilst on maintenance.
- If patients ferritin increases above 100ug/L whilst on maintenance refer patient back to clinic by e-mailing either the Lead Consultant or Nurse Specialist.
- If Hb <110-120g/L for female patient or <125-130g/L for male patient or presenting with signs/symptoms of anaemia DO NOT carry out venesection. Book patient for repeat venesection in 1 month. E-mail Lead Consultant or Nurse Specialist
- If Blood Pressure diastolic >100 or systolic <100 discuss with Dr/Registrar

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Haemochromatosis Venesection Management Plan

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