

Haemoglobinopathies - Management of Thalassaemia (Major) & Sickle Cell Disease in Pregnancy - Summary Clinical Guideline

Reference No.: UHDB/Obs/08:21/H4

Haemoglobinopathies are inherited blood disorders in which there is a haemoglobin defect resulting in an abnormal (eg sickle cell) or reduced globin formation (eg thalassaemia).

Sickle cell disease is more common in the local area. The main phenotypes which need special management are Homozygous sickle-cell disease (HbSS), Sickle-cell/HbC (HbSC) and Sickle-cell/thalassaemia (S β thal). Sickling of red cells occurs particularly to trigger factors like hypoxia, cold, acidosis, infection, dehydration and stress. Intravascular sickling leads to vaso-occlusive symptoms and tissue infarction with severe pain. They are chronically anaemic with an individual baseline haemoglobin not routinely requiring blood transfusion.

Thalassaemic disorders that matter in pregnancy are β -thalassaemia major(BTM), β -thalassaemia intermedia(BTI), HbE β thalassaemia and Haemoglobin H disease (HbH). BTM will need life-long blood transfusions and they continue with it throughout their pregnancy. The clinical course in BTI and HbH is variable. These disorders are less common locally and should be managed on a case-by-case basis.

Pregnancy in a haemoglobinopathy is associated with increased morbidity and mortality. Hence it needs to be managed as a high-risk pregnancy in the hospital jointly by a specialist Consultant Haematologist and Consultant Obstetrician with an individual pregnancy management plan that includes antenatal management, labour and delivery, and postpartum care.

Key Responsibilities and Duties

- Consultant Haematologist Clinical Lead for Haemoglobinopathy- Author, Clinical advice
- Consultant Obstetricians: Lead Obstetrician for Obstetric Haematology (preconceptual antenatal, intrapartum and postnatal care), Fetal medicine Obstetricians (prenatal diagnosis)
- Antenatal screening midwife (haemoglobinopathy screening)
- Midwife combined obstetric haematology clinic (antenatal care)
- Fetal medicine midwives (prenatal diagnosis)
- Clinical Nurse Specialist in Haemoglobinopathy (clinical advice, referral for preconceptual advice)

Communication of positive Haemoglobinopathy in Pregnancy Screening results pathway*

*Please use this flow chart in addition to the Antenatal Screening Tests guideline [OBS/03:16/H11]

Lab

Haemoglobinopathy found on antenatal screening.

Lab will authorise report so results are available on ICM and enter patient details on shared spreadsheet.

S:\Gynae\Antenatal & Newborn Screening Management\NGH screening\HAEMOGLOBINOPATHY\ANTENATAL & NEONATAL & GENERAL HB'THY\ANTENATAL\Maternity HBE referral spreadsheet.xls

Woman is a carrier for a haemoglobinopathy

Woman has a haemoglobinopathy of uncertain significance.

Enter **Maybe** on Significant Maternal Hb'thy column on spread sheet.
Inform Consultant Haematologist by email.

Woman has a clinically significant haemoglobinopathy.

For example HbSS

Enter **YES** on Significant Maternal Hb'thy column on spread sheet.
Inform Consultant Haematologist by phone & email.

- **Phone result to Antenatal Screening Coordinator (ANSC) / cover midwife in order of preference:**
- Indicate clearly if the woman has a significant haemoglobinopathy or condition of uncertain significance.

1. Phone ANSC office **89924** &/or mobile phone **07585 966169** if no answer leave voicemail
2. Phone cover ANSC (SpMW HIV) mobile phone **07799 337621** if no answer leave voicemail
3. Phone Fetal Medicine **89796 / 85409 / 89797** if no answer email as below:
4. Email to dhft.antenatalandnewbornscreeningRDH@nhs.net & Flag as **! Urgent**.

- Leave a note to check Haem spread sheet next day to ensure message has been picked up
- Contact ANSC again about any patient not actioned after 3 working days & check daily thereafter
- If results have not been picked up via Haem spread sheet after 5 working days or there are any other issues, please contact Matron for Community & Antenatal Services on **89570 &/or 07788 388437**

Maternity – ANSC or cover midwife

1. Woman is a carrier of a haemoglobinopathy

2. Woman has a haemoglobinopathy of uncertain significance:

- Enter **Maybe** on Significant Maternal Hb'thy column on spread sheet
- Inform Consultant Haematologist by email.

3. Woman has a clinically significant haemoglobinopathy: For example HbSS,

HbS/beta thal or beta thal intermediary

- Enter **YES** on Significant Maternal Hb'thy column on spread sheet.
- Inform Consultant Haematologist by phone & email, plus inform Consultant Obstetrician (FMMC)

- Check result on ICM, review Lorenzo Maternity & Obstetric notes – complete pink SC&T record sheet
- Phone woman to book ANCHM counselling apt to inform woman of result within 3 working days, ask her to bring father of baby
- Offer & (if consents) undertake screening of baby's father ASAP – chase result next day (if declined / unavailable discuss options)
- If normal phone result out OR if abnormal – recall couple for further counselling & discuss / offer PND with FMMC

Refer to NHS SC&T flow chart & table regarding risk assessment for couples

Contact Consultant Haematologist for advice & guidance on how this haemoglobinopathy is likely to behave in pregnancy

**No significant risk to woman during pregnancy.
No further action required.**

Potential risk to woman during pregnancy

Refer URGENTLY for Consultant Obstetrician-led care (with Fetal Medicine Specialism) & to Combined Obstetric Haematology Clinic

Suitable for printing to guide individual patient management but not for storage. Review Dec. July 2024