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).

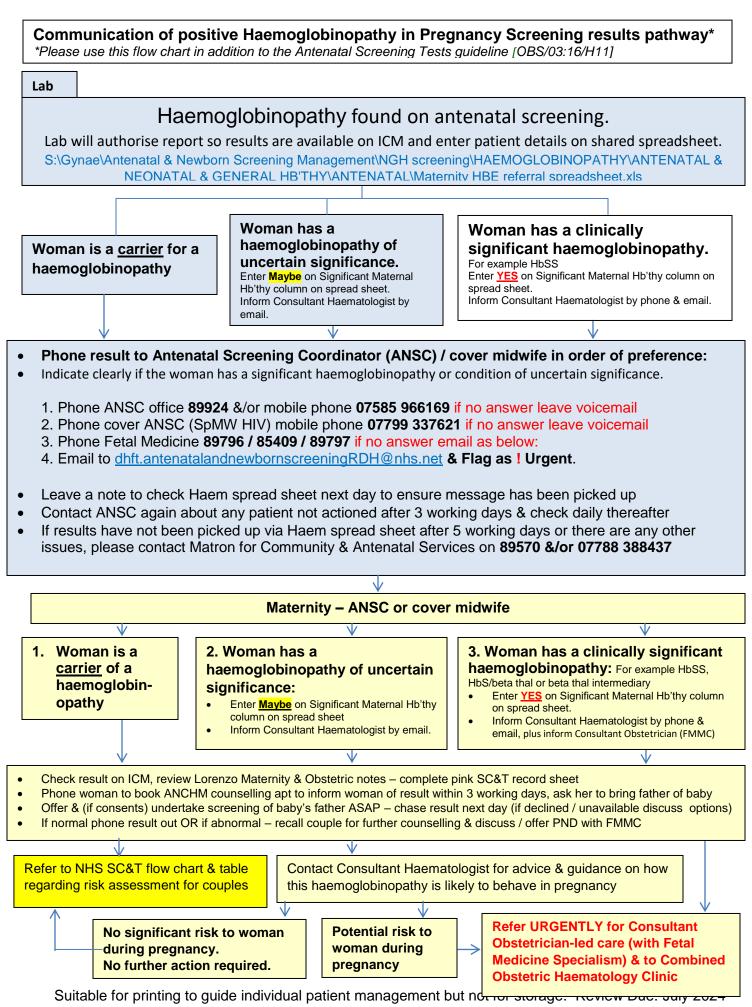
Sickle cell disease is more common in the local area. The main phenotypes which need special management are Homozygous sickle-cell disease (HbSS), Sickle-cell/HbC (HbSC) and Sickle-cell/thalassaemia (S βthal). Sickling of red cells occurs particularly to trigger factors like hypoxia, cold, acidosis, 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)



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