

## CHAPTER 30 Haemoglobinopathies in Pregnancy

Chapter 30 History					
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## 30.A Introduction

### 30.A.1 Standard Statement

Between 1991 and 1993, data was collected on Sickle Cell Disease in pregnancy from 22 hospitals. The information showed that there is a maternal mortality rate of 3.3% and a perinatal mortality figure of 6%. The increased risk, (4-5 times) of mortality and morbidity for the mother and baby demonstrates the need for clear, consistent, evidence based guidelines for all health professionals responsible for care of patients with a sickle cell disorder (SCD). Antenatal care for women with SCD should be managed at a SCD specialist centre by a multi-disciplinary team, including an obstetrician, haematologist and an anaesthetist or by a haematologist and obstetrician with expertise in SCD in a unit able to manage high risk pregnancies.

This document outlines the principles and guidelines for the care of pregnant women with Sickle Cell Disorders including HbSS, HbSC, HbSBeta thal. Hb S/O-Arab.

**For background information on Haemoglobinopathies please see [Appendix 1](#)**

Planning a pregnancy:

Primary care physicians have a key role in pre-conceptual screening, including the provision of contraceptive advice. Women with SCD should receive general pre-conceptual care (which is given to all women) and also additional advice about vaccinations, medications and crisis avoidance.

**The following is required:**

- Referral to Dr Elizabeth Rhodes and Julia Sikorska, Consultant Haematologist and Clinical lead for haemoglobinopathies in order to receive pre-conceptual counselling regarding pregnancy including the risks for mother and baby.
- Medical optimisation with organ assessment and optimal iron chelation as necessary.
- Stopping drugs which are contra-indicated during pregnancy: (Hydroxycarbamide, Chelation drugs, Hydroxyurea, ACE inhibitors, Tramadol, NSAIDS).
- A vaccination status review (Ensure Hepatitis B, Rubella and Pneumovax vaccines are up to date (within the last 5 years) that the woman has the annual influenza vaccination and that she has had haemophilus and meningococcus vaccinations.
- A health review- Taking note of transfusion history including any top up blood transfusion, elective or emergency exchange transfusions, evidence of iron overload and presence of red cell antibodies. An extended red cell phenotype should be done at first contact.
- Screening information available from any previous pregnancies. It is important to ascertain what the woman and partner already know as well. It is also important to use the information the counsellor has already, as they may have counselled the family previously.
- Counselling- The baby's father and mother are offered counselling and the father is screened for his haemoglobinopathies status.
- Early antenatal care. The woman will be under the care of the Haemaglobinopathy Team Consultant. Consultant clinics are held every Monday.
- Confirmation that the woman is taking Folic supplements 5mg daily.
- Confirmation that the woman is taking Penicillin prophylaxis 250 mg BD. If allergic to Penicillin use Erythromycin 250 mg BD (prophylactic dose).

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### 30.A.2 Antenatal care

#### Aims

- To ensure that women with SCD receive the highest standard of care during pregnancy, labour & post-partum, and to ensure the best possible outcome for mother and baby.
- To ensure that the delivery of quality care is consistent and evidence based.
- To support health professionals responsible for provision of care.

### 30.A.3 Antenatal Screening

- Written information (Screening Tests for You and Your Baby) is sent to the woman prior to her appointment. If this has not been received, it is offered again at the first appointment.
- All women are offered screening for haemoglobinopathies as part of their booking bloods as per Antenatal universal screening programme. Ideally a conclusive result is available by 10 weeks. The sample is taken together with the other booking bloods regardless of whether she has been screened before. (See Antenatal Care guideline).
- If the pregnancy has been achieved using a donor egg then the screening results for the woman will not be informative. The baby's biological father must be tested and if screen positive, the report must recommend that the fertility clinic is contacted to obtain the mother's haemoglobinopathy results.
- If the woman is known to be a carrier or affected by SCD or thalassaemia, the midwife will ask if the baby's father has been screened and, if not, offer screening for the father.
  1. Take one FBC sample (Purple bottle) and label with NHS number if known, DOB, forename, surname, as well as the routine information of date and signature. You may not have an MRN for the father. Address is required in the absence of MRN or NHS number. Note: Fathers of some nationalities can give names in a different order and the sample can become incorrectly labelled.
  2. The request form should be completed with the baby's father's details and the FBC and Sickle/Haemoglobinopathy Screen boxes ticked. Information same as above. NHS number if known, MRN number if known, DOB, forename, surname, as well as routine information of date and signature. You may not have an MRN for the father. Address is required in the absence of NHS or MRN. Note: Fathers of some nationalities can give names in a different order and the sample can become incorrectly labelled.
  3. Ethnicity is VERY important. Please ask the specific country/area and write this on the form. ([See Appendix 2](#)) Often the form/drop down menu does not allow for specific country.
  4. The woman should be referred to The Specialist Midwife/Counsellor for Haemoglobinopathies, based in the Fetal Medicine Unit on ext 3664/1911. Also inform this midwife via email of the maternal and paternal information so that the Specialist Midwife can link the couple.  
[Stghtr.antenatalScreening@nhs.net](mailto:Stghtr.antenatalScreening@nhs.net).

#### Fertility treatment

- If donor sperm has been used and the woman has a positive screening result then the report must recommend that the fertility clinic is contacted to obtain the biological father's haemoglobinopathy results.
- In the case of surrogacy, the report must recommend that the fertility clinic is

contacted to obtain haemoglobinopathy results of both biological parents.

- It is best practice to test the pregnant woman in all circumstances to ensure optimal maternal care. The report must provide the appropriate advice for the circumstances of the pregnancy.

#### Adoption

- If either biological parent has been adopted, the FAQ information may not accurately reflect the true family origins. Such cases must be treated as high risk and have full laboratory screening.

#### Bone Marrow transplants

- Where either biological parent has had a bone marrow transplant (BMT), it is likely that the results obtained will reflect the BMT donor and will not accurately represent the genetic status of the fetus. If the biological mother has had a BMT, the baby's biological father must be tested to ensure this is not a high risk pregnancy.
- If DNA confirmation of the biological mother's status is required or if the baby's biological father is post BMT and requires testing, then pre-transplant DNA or DNA obtained from hair follicles should be used.
- Please also refer the woman to the Specialist Midwife/Counsellor for haemoglobinopathies. [Stgh-tr.antenatalScreening@nhs.net](mailto:Stgh-tr.antenatalScreening@nhs.net).

#### **Late Bookers**

- Women booking late or transferring care are offered an appointment at the weekly 'late booker's slots or seen at another time, depending on individual need. All antenatal screening is discussed, and screening for haemoglobinopathies is offered as part of the booking bloods. If known to have a haemoglobinopathy variant see below.

#### **Review of Results**

- All results are sent to the team caring for the woman and reviewed by the midwife. They can also be reviewed on EPR. Abnormal results are reviewed as below. In the comments section on EPR, the clinician is made aware of whether the result has been reported to the Specialist Midwife/Counsellor. If there is no comment, the midwife should contact the service to ensure they are aware of her (telephone Ext 3664/1911 or by email to the Specialist Midwife/Counsellor for Haemoglobinopathies on either [Stgh-tr.antenatalScreening@nhs.net](mailto:Stgh-tr.antenatalScreening@nhs.net)).

### **30.A.4 Reporting of Results to Women**

#### **30.A.4.1 Normal Results**

All normal results are given to the woman by the midwife at her next midwifery follow-up appointment/ 16 weeks, if not available at the booking appointment. The result is documented in the hand-held records once it has been explained to the woman. A copy of the results is available on EPR.

#### **30.A.4.2 Inconclusive Results**

Probable Alpha Thalassaemia – results are not followed up by the haemoglobinopathy team unless indicated by the biomedical scientist. Once the result is received, the woman is called by the Specialist Midwife/Counsellor for haemoglobinopathies and the results are explained. The woman is offered a genetic counselling appointment and if screening of the baby's father is recommended then this will be indicated in the laboratory report. (Only certain partners need screening, based on the FOQ and MCH).

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**Example:**

If both parents have a sickle cell trait (HbAS), there is a 1 in 4 (25%) chance the child could inherit normal haemoglobin (Hb AA), a 2 in 4 chance (50%) the child could inherit sickle cell trait (Hb AS) and a 1 in 4 chance (25%) the child could inherit sickle cell anaemia (Hb SS).

Haemoglobin gene	A	S
A	AA	AS
S	AS	SS

For information on Family Origins and genetics please use this website: <http://www.sickle-thal.nwlh.nhs.uk/ForHealthcareProfessionals/SickleCellForHealthCareProfessionals.aspx>

Note: The Family Origins questionnaire is below in the appendices.

### **30.A.4.3 Gene Carrier/Disease**

- All abnormal antenatal haemoglobinopathy results are emailed on a daily basis by the biomedical scientist to The Specialist Midwife/Counsellor for Haemoglobinopathies and the Antenatal screening co-ordinator via Stgh-tr.antenatalScreening@nhs.net. A weekly report of abnormal results is also sent to the Specialist Midwife/Counsellor. Abnormalities include HbAS, AC, AD, AE, possible alpha zero from a high risk area, beta thalassaemia carriers etc...
- The Adult haemoglobinopathy team are also informed of antenatal results with a major haemoglobinopathy.
- The Biomedical scientist informs the Specialist Midwife/counsellor via the antenatal team generic NHS.net address. The Specialist Midwife/ Counsellor prints off the woman's demographics and haemoglobinopathy report from EPR.
- The Specialist Midwife/Counsellor for Haemoglobinopathies will call the woman and explain the result to the woman and offer a genetic counselling appointment and screening of the baby's father.
- If the genetic counselling appointment is accepted by the woman, the first action is to contact the women by phone to offer an appointment for counselling and father testing. A first appointment letter is also sent on receipt of a referral from the lab if the client is not contactable by phone.
- The woman is called and sent a text the day before the appointment to remind her of the appointment.
- If the woman declines the appointment or her partner has been screened and has a normal haemoglobin genotype, the final outcome letter is completed by Specialist Midwife/Counsellor for Haemoglobinopathies. A copy of this letter should be sent to the screening co-ordinator and the GP.
- If the woman does not attend (DNA) her appointment, the Specialist Midwife/Counsellor for Haemoglobinopathies calls the woman to find out the reason for her non-attendance. If contact is made, the woman is offered another appointment for a convenient day and time the following week. This should be followed up with an appointment letter.
- If no contact is made, Specialist Midwife/Counsellor for Haemoglobinopathies will send a second appointment letter for the following week.
- If the client DNA her second appointment, a final outcome letter is sent by the Specialist Midwife/Counsellor to the client, the client's GP and the screening

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coordinator are copied into the letter.

- If the woman attends the appointment, the outcome of the consultation is documented in their antenatal records and on Viewpoint. The final outcome letter of the consultation which includes haemoglobinopathy results of client and partner and the likely inheritance patterns of baby is sent to client, GP and the Antenatal Screening Co-ordinator.
- The Specialist Midwife/Counsellor compares the antenatal data the service has received from the lab to the antenatal data from the booking bloods data sent from the Antenatal screening coordinator to ensure women have not been missed.
- If there is no comment on the result on EPR and the Antenatal midwife has identified the woman has a result which requires following up, the midwife should contact the haemoglobinopathy team on 0208 725 3664/1911 to ensure the result is followed up.

### 30.A.5 High risk couples

High risk couples identified by the laboratory are emailed to the generic email Stgh-tr.antenatalScreening@nhs.net. The Specialist midwife will offer counselling. If the couple have been identified as high risk for sickle cell disease or Beta Thalassaemia major AND have opted for prenatal diagnosis (PND), then the Specialist midwife will refer them to the Fetal medicine consultant. They will be offered prenatal diagnostic testing. Early referral is encouraged for PND.

Women should be offered prenatal diagnosis when

- She and her baby's father are both carriers of a significant haemoglobinopathy (1 in 4 chance of the fetus being affected).
- She is a carrier of a significant haemoglobinopathy but no result from baby's father available (uncertain risk).

### Prenatal Diagnosis

Prenatal diagnosis can be performed by chorionic villus sampling or amniocentesis. This is performed in the Fetal Medicine Unit (FMU). Appointments must be made in advance by either the Specialist Midwife/Counsellor or St George's Clinical Genetics Department, as coordination with laboratories is vital.

All babies are screened for sickle cell and thalassaemia at birth as part of the newborn blood spot program. In circumstances where the parents would like to know the result sooner after birth, a capillary sample can be taken from the baby post-delivery and sent to the haemoglobinopathy screening lab at St George's. **This is a bespoke rather than routine service. See below.**

Phone Numbers:

FMU: 0208 725 3664/0080

Genetics: 0208 725 5634/1971

Specialist Midwife/Counsellor: 0208 725 3664/1911

## 30.B Pregnant women who are haemoglobinopathy gene carriers

### 30.B.1 Hb AS, Hb A $\beta$ thal, HBAE, HbAC, HbAD, HbAF

- Care for as any pregnant woman
- Clearly mark notes to indicate haemoglobinopathy carrier
- NB Possible increased risk of urinary tract infections
- Ensure partner tested
- If partners status unknown, ensure neonatal diagnosis arranged (this is routinely performed on the Universal Newborn bloodspot card)

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- Capillary testing at birth is not routinely performed. It should be discussed with parents and can be performed if parents request. Please see letter in notes from Haemoglobinopathies counsellor or the alert on E3 advising whether a heel-prick sample should be offered at birth. If indicated and agreed by the parents, the drops of blood should be collected in a paediatric FBC bottle (pale purple) by the midwife and the sample sent for electrophoresis. The request should include the parent's details and carrier status. The haemoglobinopathies Specialist Midwife/Counsellor should be informed on 0208 725 1911/3664 and will follow up the result.

### **30.B.2 Management of women affected by haemoglobinopathies**

Haemoglobinopathy disorders in pregnancy that require specialist care are:

- Sickle Cell Disease (SCD) (including HbSC, HbSS, HbSB-thal and other compound heterozygotes)
- $\beta$  thalassaemia major
- E/B thalassaemia
- Haemoglobin H disease

In the local area the most common haemoglobin disorder is Sickle Cell Disease.

### **30.B.3 Sickle cell disease**

Sickle cell disease includes women with Hb SS, Hb SC, Hb SB Thalassaemia and Hb SD. Sickle haemoglobin is a variant of the  $\beta$ -chain haemoglobin. In the deoxygenated state, sickle haemoglobin polymerises, resulting in sickling changes within the red cell. This can result in vaso-occlusion. Sickle cell patients also have chronic haemolysis and an increased rate of red cell turnover. Patients with sickle cell disease are at risk of vaso-occlusive crises such as bone pain, and chest crises. Other complications of sickle cell disease include stroke, cholecystitis, renal damage and an increased risk of infection due to splenic hypofunction.

Complications for women with SCD can occur during the antenatal period, labour, and the puerperium.

#### **30.B.3.1 Complications in pregnancy**

##### **Maternal**

- Increased risk of infection, 50% of women will develop either a chest or urine infection during pregnancy. There should be a low threshold to treat these and to encourage women to keep hydrated to prevent development of a painful crisis.
- More frequent sickle cell crises.
- Increased incidence of preterm labour, pre-eclampsia and caesarean section
- Increased risk of maternal death (approx 1%)
- Acute Chest syndrome (ACS)
- Acute anaemia

(This may be detected on routine testing. Check reticulocyte count as well. Consider non-sickle causes eg bleeding.)

- Parvovirus B19 – patients will also be reticulocytopenic. Supportive blood transfusion treatment, isolation (risk to fetus and other pregnant women)
- Sequestration – usually liver (hepatomegaly) – supportive treatment
- Acute stroke

This is a sickle complication which is not increased during pregnancy and is managed as in the non-pregnant population. Ensure urgent discussion with haematology, obstetrics and radiology. Exchange blood transfusion is the definitive treatment required.

#### **Fetal**

- Increased risk of miscarriage
- Increased risk of intra-uterine growth restriction
- Increased risk of prematurity
- Increased risk of fetal distress
- High risk of stillbirth/neonatal death (perinatal mortality 15% ie: 15 x general population)

### **30.B.3.2 Antenatal Care for known Sickle Cell Disease patients**

#### **Booking appointments**

- It is recommended that women book within 6-10 weeks with Specialist Midwife/Counsellor and team, including the Maternal medicine Consultant.
- VTE assessment.
- Flu vaccine
- Check partner's status. If HbAA, then reassure.
- First trimester diagnosis by chorionic villous sampling may be done between 11-14 weeks gestation with consent.
- Inform the woman about the extra risk of crises during pregnancy and the need to attend hospital early if crises is suspected, including seeking early medical attention if problems with nausea and vomiting, due to risk of dehydration.
- Ensure the transfusion lab has an **extended** red cell phenotype on the patient and request if not already recorded previously, antibody screening as per BCSH and NHSBT guidelines.
- Booking procedure and bloods are the same as for routine booking

#### **Medications for pregnancy Sickle Cell Disease (SCD)**

- Stop drugs which must be stopped during pregnancy: Hydroxycarbamide, chelation drugs, ACE inhibitors, tramadol, NSAIDs.
- Ensure all women are taking Folic acid 5mg daily.
- Advice to continue penicillin prophylaxis (250 mg BD or Erythromycin 250mg BD)..
- Check Hepatitis B status, immunise post-delivery if not immune.
- Daily prophylactic thromboprophylaxis, enoxaparin 40mg or Fragmin daily, (Pregnant SCD women have increased risk of PE.
- Aspirin 75mg once daily, if no contra-indications, risk IUGR.

### **30.B.4 Care of women with SCD during pregnancy**

Women with sickle cell disease are at risk of a sickle cell crisis in pregnancy, labour and the early puerperium, particularly if they become dehydrated, cold, acidotic or infected. Their care should involve specific measures to prevent these complications.

There should be involvement of:

- Consultant Haematologist,
- Consultant Obstetrician,
- Specialist Midwife/ Counsellor (who will provide antenatal care)
- Fetal Medicine Unit (FMU),
- Midwives in Day Assessment Unit/FMU,
- Anaesthetic team,
- General Practitioner, any other specialities relevant to individual patient – physiotherapist, cardiologist, renal team, etc, as below.



#### **30.B.4.1 Joint Clinic**

The high risk obstetric clinic; Monday 2pm - 5pm is held in ANC to see a Consultant obstetrician.

The Specialist Midwife/Counsellor is to check all their appointments and remind patients to attend.

Every month the hemoglobinopathies team meet to discuss patients to plan for their care. Please see appendix 3 for staff involved in the care of women with haemoglobinopathies

#### **30.B.4.2 Schedule of Antenatal Care**

All women attend a joint Haematology, Obstetric, Anaesthetic and Midwifery MDT appointment at 32 weeks where an individualised obstetric care plan will be agreed with a copy filed in the woman's obstetric notes and a copy given to the patient including the named health professional. Antenatal care will take place in Fetal Medicine Unit.

Women will be seen:

- Every 4 weeks until 34 weeks gestation (Antenatal checks should include all routine observations/care plus: FBC monthly or more frequently if clinically indicated).
- Fortnightly until 38 weeks gestation. Some patients may need more intensive monitoring depending on their individual health requirements.
- Weekly until delivery.
- Offered induction (IOL) at 39weeks gestation.

Ensure the following:

- Early dating scan or nuchal translucency scan.
- Medical review with organ assessment if not done during preconception review
- Iron supplementation is only recommended by the Haematologist. To be discussed with Haematologist.
- Anomaly scans with uterine artery Doppler at 20 weeks (Class C Recommendation).
- Four weekly scans for growth and liquor volume from 24 weeks are important
- MDT is booked between 32 and 36 weeks
- Umbilical artery Doppler if growth and liquor volume abnormal
- PET bloods at booking and every 4 weeks from 28 weeks
- MSU should be performed and sent to the lab at each visit and any urinary tract infection treated promptly.
- PCR at booking for a baseline and if clinically indicated.
- Prompt treatment of infection is vital.
- Advise to keep well hydrated.
- Advise to observe the changes of weather. The risk of crisis increases when the weather is hot or cold.

#### **30.B.4.3 Reasons for admission**

- Sick cell crisis
- Pain
- Symptomatic anaemia
- Chest pain or dyspnoea
- Pre-eclampsia
- Induction of labour
- Infection
- Labour

All pregnant Women with Sickle Cell Disease (SCD) should be seen in A+E if symptoms of crisis. On admission, the Obstetric, Anaesthetic & Haematology consultants should be informed. If out-of-hours, inform the Haematologist and Obstetrician on-call.

Take any deterioration in maternal health seriously. Have a low threshold for admission if women with SCD are unwell (eg. malaise, fever, breathlessness or pain). Make a clear differential diagnosis and plan and discuss with Consultant Haematologist.

Women having crises will generally be admitted to Delivery Suite HDU, depending on the clinical situation. If <20 weeks, women with haematological problems should be managed on the Golden Smith ward or Ruth Myles Unit and those with obstetric problems (threatened or miscarriage) in the gynaecology ward.

### 30.B.5 Pain

**ACUTE PAIN:** 25-50% of women will have at least one painful crisis during their pregnancy. - says this but then says admit!

- All pregnant women with SCD should be managed on haematology ward if crisis or unwell.
- If >20 weeks, in labour or any obstetric condition eg PV bleed, PET etc, then woman should be managed on Delivery Suite.
- Treatment of acute pain in pregnancy should follow the same guidelines as for non-pregnant women with the caveat of **avoiding NSAIDs** between 12-28 weeks or if there is proteinuria.
- Rapid assessment and appropriate analgesia should be given within 30 mins (NO PETHIDINE), with guidance from individual patient protocol if one available. Adequacy of analgesia should be assessed at 1 hour
- Parenteral opioids should be subcutaneous not intramuscular, laxatives and antiemetic as adjuncts
- Daily review by both Haematology and Obstetric teams.
- Fluids, oxygen, VTE prophylaxis, +/- antibiotic.
- Hypoxia is a significant sign and must be discussed urgently.
- Do not hyperhydrate if concerns of acute chest syndrome or PET
- Low threshold for treatment of infection.

### 30.B.6 Sickle cell crisis

#### 30.B.6.1 Precipitating factors

- Infection
- Fever
- Pre-eclampsia
- Cold
- Acidosis
- Dehydration
- Prolonged labour
- Operative delivery

#### 30.B.6.2 Management

- All in-patients with SCD should be reviewed daily by both the Obstetric, Anaesthetic and the Haematology SpR.

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- Any sign of deterioration in maternal health should be discussed with responsible physician.
- Timing and mode of delivery in relation to individual patient's health needs should be considered.
- Consider thrombo-prophylaxis in discussion with Consultant Obstetrician and Haematologist.

### **Management of the sickle cell disease crisis**

- Keep warm
- Maintain hydration. 3 litres/24 hours (Dextrose saline 1L over 8 hours). Encourage oral fluids. If unable to maintain intake give IV fluids to reduce risk of dehydration. Maintain strict fluid balance chart.
- Encourage normal diet (eating and drinking if not in labour).
- Hourly observations of Temperature, BP, Resps and pulse oximetry. Document observations on MEWS chart/HDU chart.
- 1. If febrile  $>37.5^{\circ}\text{C}$  take blood cultures and MSU and consider antibiotics
- 2. If  $\text{O}_2$  sats  $<93\%$  on air – take ABGs and get urgent medical/haematology opinion
- 3. If  $\text{PO}_2 < 9\text{kPa}$  on air – discuss need for urgent exchange transfusion with haematology team. Humidified  $\text{O}_2$  41/min if sats  $< 92\%$ :
- Pain relief (May be regular, pm, parenteral or PCA-Morphine-Diamorphine) **Avoid NSAIDs.**
- Infection screen (FBC, CRP, U&E's, LFT's)
- Physiotherapy if evidence of chest complications
- Daily CTG
- Daily urinalysis

### **30.B.7 Acute Chest Syndrome (ACS)**

This is the second most common sickle complication in pregnancy with 7-20% of patients suffering from this. It must be considered in all patients presenting with chest pain or difficulty in breathing. Clinical presentation of ACS includes tachypnoea, chest pain, lung infiltrates seen on CXR, hypoxia and often fever. It is often difficult to clinically differentiate initially from pneumonia or a PE.

- Urgent Haematology and Obstetric review
- May need a critical care review / admission
- $\text{O}_2$  therapy – may benefit from NIPV/CPAP
- Fluids – care not to hyperhydrate
- Transfusion – usually exchange transfusion is the definitive treatment
- Fragmin and antibiotics as clinically dictated

**Role of Transfusion:** There is no good evidence for routine prophylactic transfusions in pregnancy, except perhaps in the case of twin pregnancies where it should be considered.

- Top up or exchange transfusions must be discussed with Haematologists and are usually used to treat sickle complications
- Long term indications include – severe anaemia, recurrent crises, those already on a programme, those who have stopped hydroxycarbamide and are suffering from crises in pregnancy
- Extended phenotype, CMV negative blood should be used.

A randomised trial comparing prophylactic transfusion versus need based transfusion showed no difference in fetal outcome. Note that multiple transfusions increase the risk of

blood borne infections, allo-immunization and increase the need for hospital admissions. Therefore, transfusion should be reserved for symptomatic patients who are unresponsive to conservative management<sup>7</sup> (A)

Possible indications for transfusion include:

1. Hb <7g/dl
2. Recurrent crisis
3. Previous poor obstetric history
4. Patients on hydroxycarbamide pre-conception

ALL TRANSFUSIONS MUST BE DISCUSSED WITH THE HAEMATOLOGY TEAM  
All blood must have full Rhesus and Kell phenotyping

### **30.B.8 Transfusion Management**

#### **Transfusion (top up and/or partial exchange)**

- Request form for blood must be clearly marked, indicating patient has SCD.
- Check patient's ID against blood ID as per protocol.
- Observe patient for sign of incompatibility (rash, loin pain, rigors)  
1 hourly temperature, pulse and oximetry during transfusion.
- Strict fluid balance chart
- Medications as prescribed (eg Diuretics)
- In view of risk of iron overload, women with SCD tend to have exchange blood transfusion. Check the regime of transfusion for each patient.

### **30.B.9 Labour and Complications**

#### **30.B.9.1 Infections**

- 4 hourly temperature, respiratory rate, pulse and BP
- IV access and bloods (U&E's, LFT's)
- IV hydration and antibiotics
- Temp >37.5°C, take blood cultures and MSU

**Pre-eclampsia** – As per usual management of PET

#### **30.B.9.2 Labour**

- **Spontaneous rupture of membranes (SROM)**

Women with Sickle Cell Disease are advised re early induction and avoidance of multiple VE's due to increased risk of infection.

- **Induction of labour – Regime as per protocol**
- **Premature labour/Tocolysis – as per protocol**

Take any deterioration in maternal health seriously.

If in doubt, contact Dr. Ingrid Watt-Coote on pager SG903/ext 1924 or the Haematology consultant, Dr Elizabeth Rhodes/ Dr. Julia Sikorska on pager SG298 /ext 0885 or mobiles via switchboard)

#### **Fetal Surveillance**

- If a woman with sickle cell disease reports reduced fetal movement take this seriously. (The risk of IUGR is increased). She should be referred to Day Assessment Unit immediately.
- An out of hours CTG must be performed. She should be asked to attend Delivery Suite immediately if 'out of hours' and attend Day Assessment Unit at 9am on the next working day.

### Labour and delivery

Women with sickle cell anaemia are at risk of sickle cell crisis in labour and in the early puerperium. Risk is increased if dehydrated, acidotic, stressed or in the presence of an infection. Care should involve specific measures to reduce risk.

Most patients will have a pregnancy plan agreed upon at their third trimester MDT

- Patients with SCD who have normal growth scans can be offered elective births by induction of labour (IOL) or caesarean section (CS) after 37 weeks and before 40 weeks.
- Vaginal delivery is not contraindicated.
- These patients should be cared for on the obstetric HDU.
- Positioning during labour may need to be considered if patient suffers from hip pain / avascular necrosis or has hip replacements.
- Inform haematology consultant, on-call obstetric consultant and consultant anaesthetist on call once patient is admitted or in labour.
- Prolonged labour (>12 hours) should be avoided, with early recourse to caesarean section as increased stress may trigger crisis.
- If a painful crisis occurs give morphine/diamorphine.
- Bloods on admission: FBC, G&S, Ab screen and save and HbS%. Ensure blood bank know patient has sickle cell disease.
- Continuous fetal monitoring
- Hourly temp, pulse, RR and blood pressure
- Pulse oximetry- If saturations less than 92%, do ABGs and give O2 4/min.
- If evidence of chest complications, ask for physiotherapist input and inform obstetric team/haematology consultant/SpR.
- Keep well hydrated – IV access 1 litre over 8 hours with a strict fluid balance chart.

Painful sickle crises during labour should be treated as per antenatal guidelines

- Analgesia
- Opioids can be used, (**avoid pethidine, risk of seizure**).
- An epidural is beneficial for these patients **in any case**, not only if they have a crisis, therefore this should be the preferred method of pain relief in labour if no contraindications.
- Regional anaesthesia preferred for CS

**Note** paediatricians must be told about the opioid dosing and duration during pregnancy as the neonate may need to be admitted and monitored for signs of withdrawal

### 30.B.10 Care of baby

Inform the Neonatologist when woman is in labour and give history of medication. Babies may need close monitoring, particularly those whose mothers have been on considerable amount of opioids in pregnancy. These babies also need follow up by the neonatologist. The baby will need the new born blood spot on day 5 tests for sickle. A sample additional to this is only required for thalassaemia. A capillary blood sample should be done to check status if mother has requested. All baby capillary samples sent to the lab **MUST** have the demographics of both parents and their Hb status written on the form accompanying with the sample. The baby sample **MUST** be labelled appropriately according to the Screening labelling criteria.

### 30.B.11 Postnatal care

Post-delivery, women with sickle cell disease should be admitted to HDU.

Requirements include:

- 24 hours on obstetric HDU with observation – women are still at risk of sickle crisis.
- In treatment of sickle crisis NSAIDs are safe post-partum. IM/SC pain control:
- Morphine sulphate 5-10mg in at 2-4 hourly intervals.
- Review by obstetric, anaesthetic and haematology teams.
- Anticipate the need for laxatives and antiemetics with opiate analgesia.
- Keep warm
- Continue hydration: 1 litre 8 hourly for 24 hours.
- Pulse oximetry: If O<sub>2</sub> sats<92%, ABGS and give O<sub>2</sub> and seek urgent haematology and anaesthetic review.
- Chest x-ray if any chest pain or low saturation, because of risk of acute chest syndrome.
- If pyrexial – blood cultures and MSU.
- Prophylactic antibiotics (oral metronidazole 400mgs tds and cefalexin 500mg) for 5 days or Augmentin (Co-Amoxiclav) 625mg for 5 days
- Day 3 – FBC, electrophoresis if regularly transfused. Serum bilirubin.
- Early mobilisation
- Anti-embolic stockings
- LMWH – for 7 days post discharge if VD, 6 weeks if CS, or if has other risk factors for VTE identified

#### 30.B.11.1 Ongoing care

No form of contraception is completely contraindicated in patients with SCD. The progesterone only pill, depoprovera and the levonorgestrel intrauterine device (Mirena) are often preferred over oestrogen containing contraceptives and copper intrauterine devices used as second line options.

The potential increased risk of VTE with oestrogen cannot be clearly confirmed with the evidence available and so in view of this and the small infection risk with the copper IUD, the benefit of these options outweigh the disadvantages. Continue on prophylactic penicillin V (or erythromycin if allergic to penicillin) and folic acid.

#### Follow-up appts:

- P/N appointment at GP's 6/52
- Haematology appointment organised by haematology team prior to discharge.

### 30.B.12 Pregnancy with Thalassaemia

#### Introduction

- Women with thalassaemia major often have reduced fertility, because of this and the rarity of the disease, there are likely to be only small numbers of women who become pregnant and deliver at St Georges Healthcare NHS Trust. Those who do become pregnant may well have required fertility treatment.
- Ideally these patients should have in-depth preconception discussions and investigations due to the complexity of the disease, its complications and the medications involved (many of which are embryopathic).
- Pregnancy with thalassaemia major results in risks to both the mother (such as cardiomyopathy, arrhythmias, worsening osteoporosis, worsening siderosis as chelation is halted) and to the fetus (prematurity and growth restriction if ovulation

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induction has occurred and increased risks of fetal anomaly if the mother is diabetic).

An MDT approach to these patients during this time is essential.

### **30.B.12.1 Preconception Care Can**

- If failing to conceive naturally, haematologists will facilitate early referral to fertility specialists.
- Preconception meeting and discussion to plan pregnancy, ensure haematology review (MDT).
- Partner screening and counselling (as per NHS sickle cell and thalassaemia screening guidelines) – If high risk partnerships, access to discussion re preimplantation genetic diagnosis should be available. Some partners will not require screening, if it is required then this will be documented in the laboratory report.
- Early booking once pregnant.
- Viral screening
- Vaccination update
- Bisphosphonates, deferiprone, deferasirox, ACEi need to be stopped 3 months prior to conception.
- Folic acid, vitamin D measurement and supplementation, calcium supplementation
- Oral glucose tolerance test, if already diabetic then review with endocrinologists for optimisation of diabetes.

### **30.B.12.2 Thalassaemia Major**

- If patients conceive spontaneously without preconception care then care as above as well as the following:
- Cardiac review with T2\* weighted MRI of heart and liver.
- Optimisation of iron loading.
- Medication review.
- If having ovulation induction desferal can be continued until induction.
- T2\* weighted MRI if no recent imaging
- Regular anomaly scans if patient was taking deferiprone and let FMU know.

### **NO IRON SUPPLEMENTATION UNLESS DOCUMENTED IRON DEFICIENCY AND DISCUSSED WITH HAEMATOLOGY**

#### **Antenatal Care**

- MDT management
- Cardiac review at 28 weeks
- Red cell antibody screen and follow up as per BCSH / NHSBT guidelines
- DM monitoring
- Transfusion programme
- Pre transfusion Hb target 10g/dL
- Transfusion requirements likely to increase during pregnancy
- Regular growth scans, if small growth may need to increase transfusion requirements.

If known to have cardiac iron loading pre pregnancy then discussion between Haematologists, Obstetricians and Cardiologists as to whether to recommend low dose

desferal during middle pregnancy due to risk of cardiac decompensation.  
If there is severe liver loading pre pregnancy then there needs to be a formal discussion with haematologists re risks and benefits of chelation whilst pregnant.

#### **Intrapartum Care**

- Mode of delivery is guided by obstetric issues, vaginal delivery is not usually contraindicated unless there are skeletal concerns.
- Monitor Hb closely after 36 weeks. A Hb > 8g/dL should be sufficient for delivery.
- Group and Save should be sent once in labour
- Inform haematology when patient is in labour
- Strict management of diabetes

#### **Postpartum**

- VTE prophylaxis
- Desferrioxamine should be started immediately post-delivery and is not contraindicated whilst breast feeding.
- Deferiprone **IS** contraindicated during breast feeding.

#### **30.B.12.3 Thalassaemia Intermedia**

- Monthly haematology review of FBC as may need to start transfusion program
  - Regular growth scans, if reduced growth seen then consider transfusion program
- For all thalassaemia patients an individualised intrapartum plan should be drawn up in the third trimester between haematology, obstetrics, anaesthetists, cardiologists and the patient.

#### **Monitoring compliance**

Monitoring will take place by monthly audit by the Antenatal Screening Coordinator.

#### **30.B.13 Staff**

##### **Staff involved in the care of women with Haemoglobinopathies**

Consultant Maternal Medicine:	Dr Ingrid Watt-Coote ext 0071/1924
Obstetric Registrar:	Dr. Dagmar Kruger ext 4570
Consultant Haematologist:	Dr Muriel Shannon ext 1115
Consultant Haematologist	Dr Julia Sikorska ext 0885
Consultant Haematologist:	Dr Elizabeth Rhodes ext 0885
Specialist Midwife/Counsellor:	Joyce Adu-Amankwah ext. 3664/1911
Sickle Clinical Nurse Specialist:	Carol Rose
FMU Manager:	Sandra Linton ext. 0021/3664

Others who may also be involved are:

- Consultant Neonatologist
- Consultant Anaesthetist
- Cardiologist
- Nephrologist
- FMU Sonographer
- Midwives in FMU/DAU ext 3664/0863

#### **30.C References**

Rhodes E (2012) Sickle Cell & Thalassaemia Adult Service Clinical Guidelines accessed via intranet.  
Go to Units & Departments →Haematology→Haematology Disorders→Sickle Cell & Thalassaemia clinical guidelines

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British Committee for Standards in Haematology (2009) Significant haemoglobinopathies: guidelines for screening and diagnosis

National Institute for Health and Clinical Excellence. (2008). Antenatal care: Routine care for the healthy pregnant woman. London: NICE. Available at: [www.nice.org.uk](http://www.nice.org.uk)

Howard RJ, Tuck SM, Pearson TC (1995) Pregnancy in sickle cell disease in the UK: results of a multi-centre survey of the effects of prophylactic blood transfusion on maternal and fetal outcome. Br J Obstet Gynaecol 102:947 –51

Howard RJ (1996) Management of sickling conditions in pregnancy. Br J Hospital Medicine 56-1:7-10  
Notes from presentation by Susan Tuck (Consultant Obstetrician) on Pregnancy in Sickle Cell Disease at RSM (Nov 2001)

Sickle Cell Society (2008) Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK  
Collaborative Low-dose Aspirin Study in Pregnancy (CLASP) Collaborative Group. A randomised trial of low-dose aspirin for prevention and treatment of pre-eclampsia among 9364 pregnant women. Lancet 1994; 343

Oteng-Ntim E; Chase A R; Howard J; Anionwu E N (2006) Sickle Cell Disease in Pregnancy

### **30.D Appendices**

### 30.D.1 Appendix 1 Haemoglobinopathies Background Information

#### What are the Haemoglobinopathies?

Haemoglobinopathies refers to a range of genetically inherited disorders of red blood cell haemoglobin and includes sickle cell disease and the thalassaemias. Sickle cell disease and beta thalassaemia major are two of the commonest forms of these genetically inherited disorders.

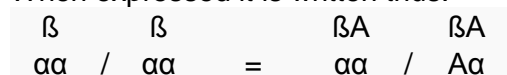
They occur most commonly in populations whose ancestors come from Africa, Asia, Mediterranean Islands, Middle, Far East Caribbean, South America and North America. Alpha thalassaemia is most common in South East Asia, Hong Kong and China.

An estimated 8,000-10,000 people with sickle cell disease and 600 with thalassaemia major live in the UK. Worldwide  $\alpha$  thalassaemia carrier states are commoner than  $\beta$  thalassaemia carrier states.

#### How the Haemoglobin Type Results in Sickle Cell Disease and Thalassaemia?

The type of haemoglobin that an individual has is determined by the haemoglobin gene that they have inherited from both their parents. Haemoglobin is the component of the red blood cell that carries oxygen around the body and each individual inherits one haemoglobin gene from each parent. The normal adult haemoglobin which predominates in individuals over the age of one year is called haemoglobin 'A' (Hb  $\alpha$ A), commonly written (HbA). It is made up of a pair of alpha and a pair of beta chains. Synthesis (production) of the beta chain is controlled by the beta globin gene located on chromosome 11, whilst production of alpha chains is controlled by the alpha globin gene located on chromosome 16. The beta gene is expressed once on each of chromosome 11 but the alpha gene is expressed twice on each of chromosome 16 and therefore synthesises a double amount of alpha chains.

When expressed it is written thus:



Where an individual has inherited two normal adult haemoglobins, one from each parent they have Hb $\alpha$ A $\alpha$ A, commonly written (HbAA). Sickle cell disease (SCD) is the family of haemoglobin disorders in which both Hb  $\alpha$  chains are abnormal, with one or both of the Hb  $\alpha$  chain genes having the sickle mutation ( $\alpha$ S). The homozygote state is HbSS, sickle cell anaemia, and a variety of compound heterozygotes exist. Thalassaemias are a group of disorders caused by the underproduction of one or more of the chains that make up the haemoglobin molecule. Hundreds of different mutations of the  $\alpha$ - and  $\beta$ - globin genes cause the clinically important thalassaemias.

#### What is a trait/carrier condition?

Where an individual has inherited one normal beta globin gene and one abnormal beta globin gene the individual has a carrier state or trait condition, which is usually benign and does not give rise to clinical symptoms or illness. For example, sickle cell trait (HbAS) and beta thalassaemia trait (HbA $\alpha$ Thal), however, carrier states have genetic implications for an individual's offspring because if their partner is also a carrier there is a one in four chance that their children can inherit a disease state.

#### How do we get sickle cell or thalassaemia syndrome?

A person can only get sickle cell disease or thalassaemia from their parents, as they can only be inherited through the genes. Each parent is born with two haemoglobin genes. Each time a couple is expecting a child, the child will have one haemoglobin gene from each parent, to

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be able to make their own haemoglobin genes. In each pregnancy there are always FOUR POSSIBLE combinations.

For example, if both parents have the usual most common combination of haemoglobin genes (HbAA) this couples children have a 100% chance of inheriting the usual combination of haemoglobins. None of their children will inherit a sickle cell or thalassaemia syndrome.

When both parents have trait conditions, where they have one normal haemoglobin gene and one abnormal haemoglobin gene such as sickle trait (HbAS) or  $\alpha$  thalassaemia trait (HbA $\alpha$ Thal) there is a chance that their children will inherit the abnormal haemoglobin gene from either or both of them. Couples of carriers have a one in four (25%) chance in each pregnancy of having a child with the disease; a one in two (50%) chance of having a child with the trait condition; and a one in four (25%) chance that the child will inherit a normal gene from both parents, and so will be completely normal.

### **What is Sickle Cell Disease?**

Sickle Cell Disease (SCD) is the term used to describe a group of inherited disorders characterised by the production of an abnormal 'sickle' haemoglobin. The sickle point mutation of the beta gene affects the quality of the beta chain produced. The most common abnormal sickle haemoglobin is sickle cell anaemia (Hb  $\alpha$ S $\alpha$ S), commonly written (HbSS). In this instance an abnormal 'sickle' haemoglobin gene has been inherited from both parents. Other common types include sickle haemoglobin C disease (Hb  $\alpha$ S $\alpha$ C) commonly written (Hb SC) and sickle beta thalassaemia (Hb  $\alpha$ S $\alpha$ Thal), commonly written as (HbS $\alpha$ Thal). Less common types include sickle haemoglobin D disease (HbSD) and sickle haemoglobin E disease (HbSE).

In SCD there is a sickle-shaped abnormality of the red blood cells. The sickle haemoglobin acquires a crystal-like formation when exposed to low oxygen tension. The oxygen in veins is low enough to cause this change and consequently the cells containing sickle haemoglobin become deformed, rigid and sickle shaped in the venous circulation. These deformed sickle shaped cells become lodged in the small blood vessels and slow or obstruct the flow of blood to the surrounding tissues causing infarction (death of the tissue). The infarction causes painful crises, known as sickle cell or vaso-occlusive crises, the hallmark of sickle cell disease. In addition the sickled cells have only a short life span due to being constantly damaged when trying to flow through the blood vessels which results in a chronic haemolytic anaemia. There is also an increased susceptibility to infections. Sufferers can be mildly, moderately or severely affected, and their pattern of pain and other problems can vary over time.

When people have sickle trait condition, they have both HbA and HbS in their blood cells. People with sickle trait are mostly well and do not have anaemia (Haemoglobinopathies registry NHS).

### 30.D.2 Appendix 2 Family Origin Questionnaire

ANC BOOKING REQUEST FORM St George's University Hospitals NHS Foundation Trust

Place labelled specimens in bag, remove protective strip, fold flap onto bag and seal firmly.

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<b>FOR LAB USE ONLY</b>		<small>Phlebotomist</small>	<b>USE BALLPOINT PEN</b>																					
		<small>WHEN REQUIRED PLACE HIGH RISK STICKER HERE ON ALL 8 COPIES</small>																						
		<b>Affix label to all copies</b>																						
Ward / Clinic _____		NHS No _____	HOSPITAL No. _____																					
Consultant _____		SURNAME _____																						
Dr Signature _____		FIRST NAME _____																						
Bleep/Aircall No _____		DOB _____																						
		MAIDEN NAME _____																						
Clinical details/past infections _____																								
Recent travel abroad: _____ (if so where?) _____																								
Signed _____	Print name _____	Job Title _____	Date _____	For urgent Chem. Path samples bleep 6032 Reception Ext. 6251																				
<b>HAVE YOU LABELLED THE SPECIMEN CORRECTLY?</b> <input type="checkbox"/> 10ml Bronze Topped Vacutainer (Gel: No Anticoagulant) <input type="checkbox"/> Pink Topped Vacutainer (EDTA: for Blood Transfusion/Bank only) <input type="checkbox"/> 8.5ml Gold Topped Vacutainer <input type="checkbox"/> Light Blue Topped Vacutainer (Citate) <input type="checkbox"/> Light Blue Topped Vacutainer (Citate) <input type="checkbox"/> Purple Topped Vacutainer (EDTA) <input type="checkbox"/> Purple Topped Vacutainer (EDTA) <input type="checkbox"/> Red Topped Vacutainer (No Anticoagulant) <b>Key to Specimen Requirements</b> <input type="checkbox"/> Red Topped Vacutainer (No Anticoagulant) <input type="checkbox"/> Bacterial Culture: Blue Topped Swab <input type="checkbox"/> Viral Culture: Green Topped Swab <input type="checkbox"/> Chlamydia: Chlamydia Collection Kit <input type="checkbox"/> All Others: Please Check Handbook																								
<b>Chemical Pathology</b> <input type="checkbox"/> Renal Profile <input type="checkbox"/> Bone Profile <input type="checkbox"/> Liver Profile <input type="checkbox"/> GGT <input type="checkbox"/> CK <input type="checkbox"/> Cardiac Profile <input type="checkbox"/> Glucose <input type="checkbox"/> HbA1c <input type="checkbox"/> Lipid Profile <input type="checkbox"/> CRP <input type="checkbox"/> B12 / Folate <input type="checkbox"/> Fe studies <input type="checkbox"/> Thyroid Function <input type="checkbox"/> LH / FSH <input type="checkbox"/> Other tests - please specify: _____		<b>Haematology</b> <input type="checkbox"/> ELSC (Sickle/Haemoglobinopathy screen + FBC) <input type="checkbox"/> ESR <input type="checkbox"/> Monospot <input type="checkbox"/> Reticulocytes <input type="checkbox"/> Malarial Parasites <input type="checkbox"/> Clotting Screen <input type="checkbox"/> D-Dimer <input type="checkbox"/> INR (Warfarin) <input type="checkbox"/> Thrombophilia Screen <input type="checkbox"/> PTT (Heparin) <input type="checkbox"/> Lupus Anticoagulant <input type="checkbox"/> Fibrinogen <input type="checkbox"/> Other tests - please specify: _____																						
Fasting <input type="checkbox"/> Non Fasting <input type="checkbox"/> On Thyroxine <input type="checkbox"/> Carbimazole <input type="checkbox"/> <input type="checkbox"/> Drug Assay (please specify) _____ Last dose: date ____/____/____ time (24hr) ____:____ Current dosage: _____ Frequency (day): <input type="checkbox"/> 1 <input type="checkbox"/> 2 <input type="checkbox"/> 3 <input type="checkbox"/> 4		<b>Medical Microbiology</b> <table border="1" style="width: 100%; border-collapse: collapse;"> <thead> <tr> <th></th> <th>Offered</th> <th>Accepted</th> <th>Declined</th> </tr> </thead> <tbody> <tr> <td>HIV</td> <td></td> <td></td> <td></td> </tr> <tr> <td>Hepatitis B</td> <td></td> <td></td> <td></td> </tr> <tr> <td>Syphilis</td> <td></td> <td></td> <td></td> </tr> <tr> <td>Additional test</td> <td></td> <td></td> <td></td> </tr> </tbody> </table>				Offered	Accepted	Declined	HIV				Hepatitis B				Syphilis				Additional test			
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HIV																								
Hepatitis B																								
Syphilis																								
Additional test																								

ANTENATAL BOOKING REQUEST	
<b>Family Origin Questionnaire</b> <small>Please tick ALL sections that apply to the woman and the baby's father</small>	
<b>A. AFRICAN - CARIBBEAN (BLACK)</b> Caribbean Islands Africa (excluding North Africa) Any other African or African-Caribbean family origins (please write in.....)	Woman <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> Baby's Father <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/>
<b>B. SOUTH ASIAN (ASIAN)</b> India or African-Indian Pakistan Bangladesh Sri Lanka	Woman <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> Baby's Father <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/>
<b>C. SOUTH EAST ASIAN (ASIAN)</b> China including Hong Kong, Taiwan, Singapore Thailand, Indonesia, Burma Malaysia, Vietnam, Philippines, Cambodia, Laos Any other Asian family origins (please write in.....) (e.g. Caribbean-Asian)	Woman <input type="checkbox"/> # <input type="checkbox"/> # <input type="checkbox"/> # <input type="checkbox"/> # Baby's Father <input type="checkbox"/> # <input type="checkbox"/> # <input type="checkbox"/> # <input type="checkbox"/> #
<b>D. OTHER NON-EUROPEAN (OTHER)</b> North Africa, South America etc Middle East (Saudi Arabia, Iran etc) Any other Non-European family origins (please write in.....)	Woman <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> Baby's Father <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/>
<b>E. SOUTHERN &amp; OTHER EUROPEAN (WHITE)</b> Sardinia Greece, Turkey, Cyprus, Italy, Portugal, Spain Any other Mediterranean Country Albania, Czech Republic, Poland, Romania, Russia etc.	Woman <input type="checkbox"/> # <input type="checkbox"/> # <input type="checkbox"/> # <input type="checkbox"/> # Baby's Father <input type="checkbox"/> # <input type="checkbox"/> # <input type="checkbox"/> # <input type="checkbox"/> #
<b>F*. UNITED KINGDOM (WHITE)</b> England, Scotland, N Ireland, Wales	Woman <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> Baby's Father <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/>
<b>G*. NORTHERN EUROPEAN (WHITE)</b> Austria, Belgium, Ireland, France, Germany, Netherlands Scandinavia, Switzerland etc Any other European family origins (please write in.....) (e.g. Australia, N America, South Africa)	Woman <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> Baby's Father <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/> <input type="checkbox"/>
<b>*Hb Variant Screening Requested by (F) and/or (G)</b> # Higher risk for alpha zero thalassaemia	
<b>H. DON'T KNOW</b> (incl. pregnancies with donor egg/sperm) <b>I. DECLINED TO ANSWER</b> <b>J. GESTATION AT TIME OF TEST</b> SCREENING TEST DECLINED <input type="checkbox"/> Do you want to give a reason why declined? _____	
Yes ..... No <input type="checkbox"/>	
The completion of this form is an ESSENTIAL part of NHS Screening Programme for sickle cell & thalassaemia	

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### 30.D.3 Appendix 3 Sickle cell disease Summary of Care Antenatal care

- See Antenatal care pathway for Haemoglobinopathies – **Chapter 30**
- Women need to book under Specialist Midwife/Counsellor who will arrange referral to Mulberry team
- Partner testing
- Ensure women are taking:
  - Folic acid 5mg od
  - Penicillin V prophylaxis
- Discuss and commence:
  - Aspirin 75mg od
  - Thromboprophylaxis (Straight away if any additional VTE risk factors)
  - From 28 weeks if no additional VTE risk factors
- PCR at booking and then if indicated. Monthly FBC/reticulocytes/MSU
- Regular monitoring for detection of red cell antibodies
- Serial growth scans
- MDT meeting between Maternal Medicine, Anaesthetic and Haematology teams at 34-36 weeks to finalise birth plan (IOL 38-39 weeks)

#### Intrapartum care

- See Antenatal care pathway for Haemoglobinopathies
- Plan for monitoring in labour to be finalised at MDT
- IV access, FBC, G +S, Ab screen/ HbS% and cross match 2 units blood
- Hourly monitoring of BP/HR/RR/Oxygen saturations/Temperature
- Ensure hydration/oxygenation maintained
- Keep warm
- Continuous fetal monitoring in labour

#### Postpartum care

- Check care plan made at MDT (likely to require Obstetric HDU care for a minimum of 24 hours postpartum)
- NSAIDs are safe
- Morphine sulphate prescription
- Ensure hydration/oxygenation maintained
- Obstetric/Anaesthetic and Haematology review prior to discharge
- Discuss contraception
- Prophylactic antibiotics (oral metronidazole 400mgs tds and cefalexin 500mg) for 5 days or Augmentin (Co-Amoxiclav) 625mg for 5 days
- Day 3 – FBC, electrophoresis if regularly transfused. Serum bilirubin.
- Anti-embolic stockings
- LMWH – for 7 days post discharge if VD, 6 weeks if CS, or if has other risk factors for VTE identified

#### Contact details

Consultant Haematologist with Special Interest in Sickle Cell Disease: Dr Elizabeth Rhodes  
07919184441

Haematology SpR on call: Bleep 6068

Joyce Adu-Amankwah (Specialist Midwife/Counsellor for Haemoglobinopathies) FMU ex  
3664/1911

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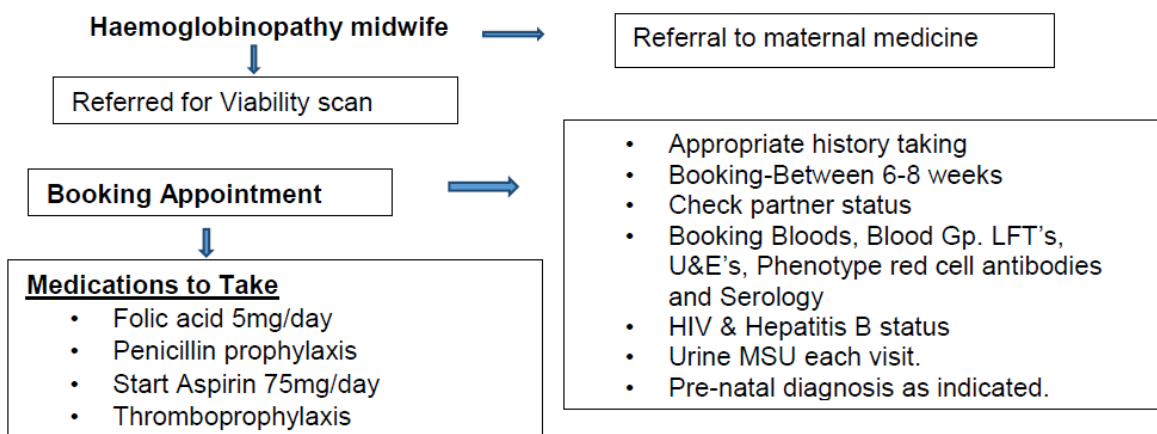
### 30.D.4 Appendix 4 Antenatal/Intrapartum and Postnatal care pathway

#### Antenatal care pathway for Haemoglobinopathies

Haemoglobinopathies are a group of disorders affecting either haemoglobin structure / haemoglobin synthesis.

##### Aim of Screening:

- To minimize delays in identification of high risk women of having an affected infant and to identify genetic carriers (1 in 4 chance of affected baby).
- To improve Co-ordination, communication & liaison between healthcare professionals.
- Once identified as at risk- Referred to Haemoglobinopathy midwife.
- The laboratory will involve the sickle & thalassemia service team at Balham regarding all the results and they would contact the mother and offer counselling/screening the baby's father or offer prenatal diagnosis (PND) if needed.



Name			
Hospital Number		Date of Birth& Age	
Pregnancy Dates EDD		Booked at SGH or elsewhere?	
No. of prev pregnancies		Details of previous pregnancies	
Sickle Diagnosis		Partner Diagnosis	
High Risk Pregnancy? (risk of affected baby)		Counselled? Actions	
Other History	Sickle History		
Any significant family history? Any significant social history? HADS score? Any mental Health concerns?	Multiple Admissions? ICU admission? Previous surgery? Acute Chest Syndrome? Stroke? Pulmonary HTN? Proteinuria?		

<b>HAEMATOLOGY AND TRANSFUSION HISTORY</b>			
Baseline Haemoglobin		Ferritin level:	
Previous Transfusions?		Regular transfusions?	
Extended Phenotype Sent? Blood bank informed of pregnancy		Antibodies Known?	
<b><u>MEDICATIONS</u></b>			<b>Actions</b>
Hydroxycarbamide?		<b>SHOULD BE STOPPED DURING PREGNANCY</b>	
Chelation?		<b>SHOULD BE STOPPED DURING PREGNANCY</b>	
Ace Inhibitors		<b>SHOULD BE STOPPED DURING PREGNANCY</b>	
Pain killers discussed		<b>NSAIDS and TRAMADOL SHOULD BE STOPPED DURING PREGNANCY (If necessary NSAIDS can be given as PRN between 14-32 wks).</b>	

**Antenatal care- Follow up clinic-With Case-Load midwife's-Ms Joyce Adu-Amankwah / Ms Sandra Linton and Obstetrician- Ms Ingrid Watt-Coote.**

**Women will be seen by the Multidisciplinary Maternal Team by 34-36 weeks**

**Obstetrician- Dr I Watt-Coote  
Haematologist-Dr E Rhodes  
Anaesthetist  
Case load midwife**

- Check all results
- Monthly FBC, Reticulocytes and MSU for Culture
- To book into MDT by 26 wks
- Red cell antibodies at booking, 28wks & 34 wks / before each transfusion.
- Every 4 weeks until 24 weeks
- Fortnightly until 32 weeks
- Then weekly until delivery
- Check MDT done, 34-36wks.

#### **Ultrasound Scans**

#### **Salient points to remember**

- Early dating scan / NT scan
- Anomaly scan with Uterine artery dopplers 20-21 wks
- Serial Growth scans
- Escalate to MDT if any concerns regarding uteroplacental insufficiency.

- Document any episodes of sickle pain, record sites, duration, severity and analgesia used.
- ECHO and Retinopathy screening to be arranged by sickle cell midwife as indicated.
- Delivery to be planned by 38 weeks gestation. Vaginal delivery could be offered and Caesarean section only for Obstetric reasons.

• Investigations	Date	Result	Action
Echo and baseline SpaO2			
BP and urine dip			
Other organ assessment as needed			
Actions from first appt <ul style="list-style-type: none"> <li>• Investigations to be ordered</li> <li>• Outstanding results</li> <li>• Referrals to other specialties</li> <li>• Community Team aware</li> <li>• Transfusion plan needed?</li> <li>• Alternatives to Hydroxyurea discussed</li> </ul>			
Pain plan discussed – analgesia requirements			
Monthly haematology appts until 24/40 then two weeks <ul style="list-style-type: none"> <li>• Check urine</li> <li>• Check Haemoglobin</li> <li>• Check Ab status</li> <li>• Check problems with sickling</li> </ul>			
Obstetric / Anaesthetic / Haem MDT at 34-36wks and pregnancy plan			
Other Issues:			