Haemoglobinopathies in Pregnancy – Guideline for Management

University Hospitals of Leicester NHS Trust

C74/2006

Contents

1. Introduction and who the guideline applies to:

This guideline is intended for the use by all medical, midwifery, ultrasonography, haematology, and pharmacy staff involved in the care of pregnant women with sickle cell disease and thalassaemia.

Related documents:

- **Sickle Cell Disease - Chronic Complications UHL Haematology Guideline** C36/2013
- **Sickle Cell and Thalassaemia (Haemoglobinopathy) Screening in Pregnancy UHL Obstetric Guideline** C58/2006
- **Sickle Cell Crisis in Adults - Subcutaneous Patient Controlled Analgesia UHL Policy** C1/2021
- **Thromboprophylaxis in Pregnancy Labour and Vaginal Delivery UHL Obstetric Guideline.pdf** C1/2017

Background

Haemoglobinopathies are conditions in which there is an inherent haemoglobin defect resulting in abnormal (e.g. sickle cell) or reduced globin formation (e.g. thalassaemia).
This guideline will cover the following haemoglobinopathies that require management in pregnancy which are:

1. Sickle Cell disease
2. Transfusion dependent and non-transfusion dependent thalassaemia

Please note: Sickle cell trait (HbAS) is a carrier state that may contribute to anaemia in pregnancy but does not cause any of the significant maternal or fetal complications outlines below. A couple who both have sickle cell trait will have a 1 in 4 (25%) risk of having a child with sickle cell disease. This would be apparent following routine haemoglobinopathy antenatal screening and patients counselled by the appropriate clinical team.

2. Sickle cell disease (SCD)

- This is a group of inherited blood disorders characterised by sickling of red blood cells during physiological stress leading to vaso-occlusion. This most commonly leads to painful crises, but can cause more serious complications.
- Patients with sickle cell disease have increased red cell turnover and a chronic haemolytic anaemia which will also be affected by the physiological changes of pregnancy.

The most common forms of sickle cell disease are:

- HbSS.
- HbSC.
- HBS β-thalassaemia.

More rarely there are other causes of sickle cell disease:

- HbSD-Punjab
- HbSE.
- HbSO-Arab.

HbSS (sickle cell anaemia) is associated with the highest risks during pregnancy but all forms of sickle cell disease should be managed based on the recommendations outlined. Maternal mortality has been reported at a rate of 1-2% (Howard et al 1995, Smith et al 1996). Close monitoring of pregnancy women in a multidisciplinary setting is crucial in order to minimise the risks of complications.

Table 1: Complications

Maternal Complications	**Foetal Complications**
Worsening anaemia	Spontaneous miscarriage
Increased risk of infections, particularly UTI and chest infection	Intrauterine growth restriction
Increased sickle cell crises, particularly in the third trimester	Perinatal mortality
Acute Chest syndrome	Pre-term delivery – iatrogenic and idiopathic
Hypertension and pre-eclampsia	Still birth
Thromboembolic disease	Opiate toxicity in the neonate
Antepartum haemorrhage	

2.1 Management:

2.1.1 Pre-pregnancy

- All women with sickle cell disease are advised to plan their pregnancy and can be offered pre-conception counselling if requested. This will include discussion of the risks related to pregnancy and review of potentially teratogenic medication. The local haemoglobinopathy sickle cell nurse will be able to coordinate this. This will allow women with any specific risks, such as those relating to red cell alloantibodies or end organ damage, the opportunity to discuss these in relation to any future pregnancy
- Hydroxycarbamide should be discontinued in both men and women planning pregnancy, at least 3 months prior to conception. This should be reviewed in regional MDT if a woman is considered to be high risk and transfusion is not feasible. If conception occurs, however, hydroxycarbamide is not in itself a reason for termination.
- ACE inhibitors and chelation therapy should also be stopped.
- Where possible, genetic screening and partner testing should also be performed, as pre-implantation genetic diagnosis can be performed in selected couples.
- Vitamin D deficiency is common in women with SCD. Regular monitoring and supplementation outside of pregnancy is recommended and this should be encouraged, given supplementation is recommended for all pregnant women.
- Vaccinations should be kept updated in line with national recommendations

2.1.2 Early Pregnancy

- Identification of a pregnant woman can occur in a number of ways. Once identified, women should be referred to the regional specialist haemoglobinopathy centre for review in a joint obstetric haematology clinic (see flowchart)
- Antenatal care will be provided by the regional specialist multidisciplinary team with regular correspondence to the local team (if applicable)
- Women will receive consultant led care in the tertiary specialist centre
- Delivery will be booked in a consultant led unit. This may be at the local centre if felt safe to do so, but all cases will be assessed on an individual basis
- An individualised care plan will be made and documented in the maternity notes and hand held notes (see appendix 1&2)
- An early booking appointment will be offered and the assessment will include;
    - **Confirmation of viability:** An early viability scan (7-9 weeks) is encouraged
    - **Review of medication:** Stop any potentially teratogenic medications. Start aspirin 150mg daily, Vitamin D 1000iu supplementation and continue folic acid 5mg OD.
    - **Ensure partner screening:** Offer pre-natal diagnosis is if appropriate (if performed, results should be available by 12+6/40 in line with screening standards).
    - **Screen for end organ damage:** If no echocardiogram or ophthalmology assessment in previous 12 months, repeat in pregnancy. Review previous urine PCR for proteinuria and baseline BP.
    - **Review vaccinations:** Review previous history and consider from 2nd trimester onwards if appropriate. Continue penicillin prophylaxis throughout pregnancy (or erythromycin if penicillin allergy).
    - **Allo-antibody screening:** Perform in line with national screening programme at booking but if previously known, ensure appropriate follow up and monitoring in place. Perform red cell phenotype if not previously done.
    - **Assessment of disease severity and pregnancy risk**: Routine prophylactic transfusion is not currently recommended but all patients should be assessed for the potential benefit based on disease severity. This will be reviewed at the next available EMSTN network meeting and should take previous pregnancy

morbidity into account (see appendix 3 for pathway). Patients on a transfusion programme prior to pregnancy should continue this throughout pregnancy. Frequency of procedures will need to be reviewed, along with iron status.

- **Review steady state levels:** To include baseline FBC, ferritin, folate, LFT, U&E, LDH, Reticulocyte and HbS% if recently transfused. Offer iron replacement if indicated.
- **VTE risk assessment:** All women will score 3 based on the diagnosis of sickle cell disease and require LMWH thromboprophylaxis from 28/40. Any additional risk factor will result in the need for thromboprophylaxis throughout pregnancy.
- **Care plan:** To include review of previous analgesia management and documentation of management in event of crisis pain (see appendix 2)
- **Education:** should pay particular attention to:
    1. Discussion of maternal and fetal risks
    2. Crisis prevention measures, including avoiding exposure to extreme temperatures, dehydration and overexertion
    3. Particular attention to persistent vomiting, which should prompt early medical advice
    4. Outline of antenatal care plan
    5. Devise and circulate individual plan of treatment for crises.

2.1.3 Blood Transfusion

Routine prophylactic transfusion is not recommended throughout pregnancy but in the event of a transfusion being required this must be discussed with the on call haematology team. Blood should be matched for extended red cell phenotype, Kell and be CMV and sickle negative (see local transfusion policy).

Prophylactic transfusion should be considered in women with:
1. Previous or current medical, obstetric or foetal problems related to SCD
2. Women previously on hydroxycarbamide due to severe disease
3. Multiple pregnancy

Top up transfusions may be required if the haemoglobin falls below the patient baseline (usually 60-80g/L in HbSS) due to aplastic or haemolytic crisis. Patients on pre-pregnancy transfusion programmes should continue throughout the pregnancy.
Transfusion should also be considered for those with acute SCD complications.

2.1.4 Ongoing antenatal care

- Women should be seen in a joint clinic routinely every 4 weeks until 28/40 and then every 2 weeks. Routine antenatal care should be delivered alongside this.
- At each antenatal visit (see checklist)
    - Assess compliance with medication
    - Assess foetal growth. The RCOG suggest regular fetal growth scans should be performed every 4 weeks from 24 weeks gestation. An individualised care plan will be made after a risk assessment by the mutli-disciplinary team. The scan interval may be based on specific fetal concerns and/or maternal concerns e.g. episode of severe crisis, exchange transfusion
    - Monitor blood pressure and urinalysis (monitor for PET and UTI)
    - Monitor FBC/Reticulocyte/LDH/LFT/ferritin/U&E
- Repeat standard ABO and red cell antibody screen at 28 weeks gestation and follow local guidelines if an antibody is detected
- Review thromboprophylaxis in line with local policy
- Offer anaesthetic assessment in the third trimester of pregnancy.

- Discussion regarding labour management and options for delivery. Prepare labour plan at 36 weeks gestation (if appropriate), with particular attention to mode of delivery. Aspirin can be reviewed at this time to assess if can be stopped prior to delivery.
- For patients referred from outside of a tertiary centre, a local antenatal appointment in the third trimester will be offered to ensure local plans for delivery are in place in the case of emergency presentation in labour

2.1.5 Labour and delivery

- If pregnancy is uncomplicated, delivery should be planned for 38 to 40 weeks gestation. Mode of delivery will be determined by obstetric factors Cross matched, appropriate blood should be available for delivery if required. Ensure FBC and G&S sample on admission.
    - Hb<80g/l should prompt consideration of red cell transfusion, but all transfusions should be discussed with the on call haematology team.
- The risk of sickle cell crisis in labour and the early puerperium is increased if the woman becomes dehydrated, acidotic or has active infection. Care needs to be directed towards preventing these by specific measures by the following:
    - Keep warm
    - Maintain good hydration: Commence intravenous fluids at a rate of 125ml/hour on admission (if safe to do so) and monitor urine output
    - Strict recording of fluid balance to avoid fluid overload.
    - Continuous pulse oximetry-if oxygen saturation is less than 95% on air, consider oxygen via a mask at 5L/minute **and escalate to clinical team**
    - Continuous CTG monitoring throughout labour.
    - Epidural analgesia is the pain relief of choice. Pethidine should not be given. If sickle pain develops, patient controlled analgesia may be required.
    - **Avoid prolonged labour** (not longer than 12 hours) with early recourse to caesarean section.

2.1.6 Postpartum Care

Baby

- Send capillary blood for screening for sickle cell status (in addition to newborn blood spot)

Mother

- Maintain adequate hydration and oxygenation as above.
- 4 hourly observations on maternal early warning score (MEOWS) for at least 24 hours post delivery
- Monitor for infection with low threshold for antibiotics. If required, continue IV antibiotics for 24 hours post-delivery then review and consider continuing antibiotics orally for 7-14 days. (see Antimicrobial Summary UHL Womens Guideline)
- Check FBC/LFT/Reticulocyte/LDH on day 1 and 3 postpartum (as a minimum). Further testing based on clinical scenario.
- Thromboprophylaxis with weight appropriate LMWH for six weeks post-partum
- Review as an inpatient by haematology team and on discharge, follow up in the joint clinic for postnatal check
- Contraception should be considered and discussed
- Encourage early mobilisation and breast feeding

2.2 Sickle Cell Crisis (see protocol in appendix 2)

- This is the most common reason for admission and occurs in 7-20% of pregnancies. Pain is the prominent feature and prompt analgesia is required.
- An agreed pain management plan for the treatment of acute pain crisis should be documented (appendix 2)
- Women may present to haematology or obstetrics and both teams should be made aware. Any woman who presents in crisis should be discussed with the on call haematologist.
    - Women 23should be admitted to haematology
    - Women ≥ 23should be admitted to obstetrics

2.2.1 Immediate Measures:

1. Give appropriate analgesia: This should be within 30 minutes of presentation as per NICE. This may include paracetamol and diclofenac , but often opiate analgesia is required (see appendix)
    a. If opiates are required, Morphine should be dosed based on booking weight:
        i. If <50kg, give 5mg morphine sulphate SC/IV
        ii. If ≥50kg, give 10mg morphine sulphate SC/IV
2. Assess response: Review after 30 minutes and give a second dose of morphine if required (provided there is no evidence of opioid toxicity). Ongoing monitoring should continue at least every 4 hours.
   Note: Respiratory rate and conscious level should be monitored every 15-30 minutes initially then every 4 hours unless specific concerns.
3. Monitor oxygen levels: If oxygen saturations are <95% on air, offer oxygen at 5l/min **and escalate to medical team. Inform haematology on call.**
4. Keep warm
5. Ensure adequate hydration: Aim for 60ml/kg/24 hours and monitor fluid balance. Give intravenous fluids if required
6. Monitor baby**:** If ≥ 24/40, monitor with CTG

2.2.2 Assessment and further management

1. Investigations: Check FBC/U&E/LFT/LDH/reticulocytes/coagulation, HbS% (if recently transfused). Consider CXR if hypoxia and/or chest sepsis suspected
2. Screen for infection: Send cultures (if appropriate) to microbiology;
3. Consider antibiotics: If features of infection (such as fever, unexplained tachycardia, focal symptoms). Stop penicillin V prophylaxis if treatment antibiotics started **and restart on completion of antibiotic course**
4. Regular pain relief: Ensure regular analgesia continued. If required, consider patient controlled analgesia. Offer incentive spirometry.
5. Reassess: Review pain relief, investigations and evidence of infection. Monitor observations in line with trust policy. **If signs of opiate toxicity (such as low RR), omit morphine and consider naloxone 100micrograms as necessary**
6. Haematology review: Inform on call haematology team for review.

2.3 Acute chest syndrome

- Life threatening complication associated with abnormal CXR and/or hypoxia due to acute sickling in lung tissue. Patients may present with ACS or this can develop as a complication of an acute painful crisis
- **Any** hypoxia (O_2 sats <95% air) or escalating oxygen requirements should be escalated to the on call haematologist

- HDU/ITU input may be required and transfusion should be considered. Consideration of an exchange transfusion will be made by the on call haematologist.

3. Thalassaemia

- Group of inherited blood disorders with abnormal formation of red blood cells
- Women may be transfusion dependent (previously called thalassaemia major) or non-transfusion dependent (thalassaemia intermedia)
    - Transfusion dependent women need their medical care optimised before pregnancy where possible as this can be associated with organ damage due to iron overload (cardiac disease, diabetes). This can lead to increased risks to the mother and safety of pregnancy should be considered. Iron chelation should be reviewed, and where possible, stopped 3 months pre- conception.
    - Non-transfusion dependent women may require transfusion support in pregnancy due to the physiological changes which occur and so should be monitored by a specialist team.

3.1 Early pregnancy

- Antenatal care will be provided by the regional specialist multidisciplinary team with regular correspondence to the local team (if applicable)
- Women will receive consultant led care in the tertiary specialist centre
- An early booking appointment should be offered and the following discussed:
    - **Confirmation of viability:** Early scan (7-9 weeks) must be performed
    - **Review of medication:** Stop any potentially teratogenic medications (such as chelation, bisphosphonates) and ensure 5mg folic acid continued
    - **Ensure partner screening:** Offer pre-natal diagnosis is if appropriate (if performed, results should be available by 12+6/40 in line with screening standards).
    - **Screen for end organ damage and assessment of pregnancy risk:** Review most recent annual review for any evidence of known abnormalities, including most recent assessment of iron status. Patients with pre-existing cardiomyopathy should be discussed with an appropriate cardiologist. If pre-existing diabetes, additional input by the diabetic team will be required.
    - **Allo-antibody screening:** Perform in line with national screening programme at booking but if previously known, ensure appropriate follow up and monitoring in place. Perform red cell phenotype if not previously done.
    - **Offer aspirin and LMWH:** If previously undergone splenectomy or platelet count >600, in addition to obstetric indications. Consider LMWH if increased thrombotic risk in line with RCOG recommendations.
    - **Review transfusion requirements:**
        - If regularly transfused, the frequency may increase in the later stage of pregnancy but this will be reviewed by the consultant haematologist. Pre-transfuison aims of >100g/l may not be possible due to the physiology of pregnancy.
        - CMV negative blood must be transfused (in addition to known requirements). It is the responsibility of the haematology team to inform blood bank of this.
        - If not-regularly transfused, inform women of the probable need for blood transfusion during pregnancy. Perform red cell phenotyping if not previously done.

3.2 Ongoing antenatal care

- See women in a joint obstetric/haematology clinic every 4 weeks until 28/40 then every 2 weeks thereafter
- Perform FBC at each visit and assess for symptoms of anaemia
- Offer close monitoring of fetal growth, RCOG suggest every 4 weeks from 24/40 – an individualised care plan will be made after risk assessment by the multi-disciplinary team.
- In exceptional circumstances, desferrioxamine chelation may be used from 20-24/40 in those women with evidence of severe iron overload and who may be at risk of cardiac decompensation. This decision must be made by a consultant haematologist following extensive multidisciplinary discussion.

3.3 Labour and delivery

- Vaginal delivery is preferred and delivery plans determined by obstetric implications
- Aim to optimise haemoglobin prior to delivery; If presents in labour, send FBC and G&S sample
- Intravenous desferrioxamine should be given to women with transfusion dependent thalassaemia in during delivery
- Offer continuous intrapartum fetal monitoring
- Active management of third stage of labour to minimise bleeding

3.4 Postpartum care

Baby

- Send cord sample for haemoglobinopathy screen if baby at risk (in addition to neonatal blood spot)

Mother

- Encourage breastfeeding and mobilisation
- Review thromboprophylaxis in line with RCOG guidelines
- Check maternal FBC day 1 postpartum to optimise post-natal haemoglobin
- Desferrioxamine can be switched to SC once 24 hour IV dose completed (excreted in breast milk but not orally absorbed). Should be chelation of choice if breast feeding continues as minimal safety data for alternatives
- Offer 6 week post natal review in obstetric haematology clinic

4. Education and Training

None

5. Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
None				

6. Supporting References

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2. Ladwig P, Murray H (2000) Sickle cell disease in pregnancy. Australia and New Zealand journal of Obstetrics and gynaecology; 40(1): 97-100.

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5. Powars DR ,Sandhu M, Niland-Weiss J,Johnson C, Bruce s, Manning PR Pregnancy in sickle cell disease. Obstetrics and Gynaecology 1986; 67:217-228.

6. Adam S (1996) Sickle cell disease in pregnancy. Caring for the pregnant women with sickle cell crisis. Professional Care of Mother and child;6(2): 34-36

7. De Swiet M MEDICAL Disorders in Obstetric practice 3rd Ed. Oxford: Blackwell Science 1995; Chapter 2: 50-57.

8. Koshy m ,Burd L ,Wallace D Moawad A Baron J(1998) Prophylactic red cell transfusions in pregnant patients with sickle cell disease. The New England Journal of Medicine; 319(22): 1447-1452.

9. RCOG Green top Guidelines 61:2011.

10. RCOG Green top guideline 66 – Management of Beta Thalassaemia in pregnancy 2014

11. Management of sickle cell disease in Pregnancy: A British Society Guideline for Haematology guideline September 2021

Related documents:

- Sickle Cell Disease - Chronic Complications UHL Haematology Guideline C36/2013
- Sickle Cell and Thalassaemia (Haemoglobinopathy) Screening in Pregnancy UHL Obstetric Guideline C58/2006
- Sickle Cell Crisis in Adults - Subcutaneous Patient Controlled Analgesia UHL Policy C1/2021
- Thromboprophylaxis in Pregnancy Labour and Vaginal Delivery UHL Obstetric Guideline.pdf C1/2017

7. Key Words

Sickle cell disease in pregnancy, sickle cell crisis, Thalassaemia, Haemoglobinopathies

The Trust recognises the diversity of the local community it serves. Our aim therefore is to provide a safe environment free from discrimination and treat all individuals fairly with dignity and appropriately according to their needs.
As part of its development, this policy and its impact on equality have been reviewed and no detriment was identified.

Contact and review details	
Guideline Lead (Name and Title) A Webster - Consultant Haematologist N Archer – Consultant obstetrician	**Executive Lead** Chief medical officer
Details of Changes made during review: Added to conduct MDT meeting pre-pregnancy when discussing discontinuation of Hydroxycarbamide Added reference to vitamin D supplementation and continuation with routine vaccinations Rationale added for prophylactic blood transfusion Added routine A/N care should be provided alongside specialist care Rationale for possible reduced standard USS intervals dependent on MDT risk assessment Review Aspirin at 36/40 SCC admission gestations changed from <24 to <23 to haematology & >23 to obstetrics (prev >24) Checklist enlarged and updated	

Appendix 1: Pregnancy in women with sickle cell disease: Checklist

Patient details (insert addressograph)

Patient diagnosis:
Baseline results: Hb Retics Sats on air
Blood group: **Red cell antibodies**: Y / N
Partner screening result:
Significant history:
Primary Haemoglobinopathy centre: ……..
Consultant Haematologist/Obstetrician in pregnancy:
……

Please note this is a checklist for basic care requirements in pregnancy
If additional risks known please refer to relevant local protocol for additional management

Obstetric Haematology visit (by gestation)	Visit requirements	Confirm completed: Sign and date
6-12/40 (first clinic visit)	• Confirm viability (increased risk of miscarriage) • Discuss information, education and advice regarding impact of SCD in pregnancy (outline both SCD and pregnancy related complications). Offer general health information and support. • Review partner screening results • Assess retinal, renal and cardiac complications. If not screened with previous 12/12, needs referral in pregnancy • Review red cell antibody screening • Baseline blood tests (FBC, Ferritin, Folate, U&E, LFT, LDH) • Start aspirin 150mg OD and review need for Vitamin D • RCOG VTE score – advise routine administration from 28/40 unless additional risk factors • Complete sickle cell crisis plan and file in notes • Discuss vaccination history • Review medication – offer routine penicillin prophylaxis in pregnancy, continue folic acid 5mg and ensure teratogenic meds stopped • Discuss and review need for regular transfusion programme • Refer to regional haemoglobinopathy MDT for discussion • Copy correspondence to local haematologist and obstetric team (where applicable)	

Ongoing review: Medical review at each visit – ensure correspondence copied to local team (if applicable)

Routinely see every 4 weeks until 28/40 then every 2 weeks until delivery

16/40	• Investigations: BP, urinalysis and MSU • Submit Anaesthetic referral	
20/40	• Foetal anomaly scan • Investigations: Bloods (FBC), BP, Urinalysis and MSU	
24/40	• Growth scan • Investigations: Bloods (FBC), BP, urinalysis and MSU	
28/40	• Growth scan • Start LMWH prophylaxis unless contraindicated (if not previously done) • Consider glucose tolerance test • Investigations: Bloods (FBC, G&S), BP, Urinalysis and MSU	
30/40 **To be completed at local hospital/midwife if applicable**	• Investigations: BP, urinalysis and MSU	
32/40	• Growth scan • Arrange anaesthetic review • Investigations: Bloods (FBC), BP, Urinalysis and MSU	
34/40	• Investigations: BP, urinalysis and MSU • Discuss delivery and provisional delivery plan	
36/40	• Growth scan • Investigations: Bloods (FBC), BP, Urinalysis and MSU • Arrange delivery (if not previously done)	
38/40 (if not delivered)	• Investigations: BP, urinalysis and MSU • Arrange delivery (if not previously done)	

Copies of this document should be available in:

Handheld notes

Hospital medical notes

Maternity medical notes

If any concerns, please contact consultant haematologist or obstetrician in pregnancy as outlined above

Reviewed January 2022

Appendix 2: Management of Vaso-occlusive protocol

EMSTN
East Midlands Sickle Cell and Thalassaemia Network

Management plan for sickle cell crisis in pregnancy

Patient details (insert addressograph)

Patient diagnosis:

Baseline results: Hb Retics Sats on air

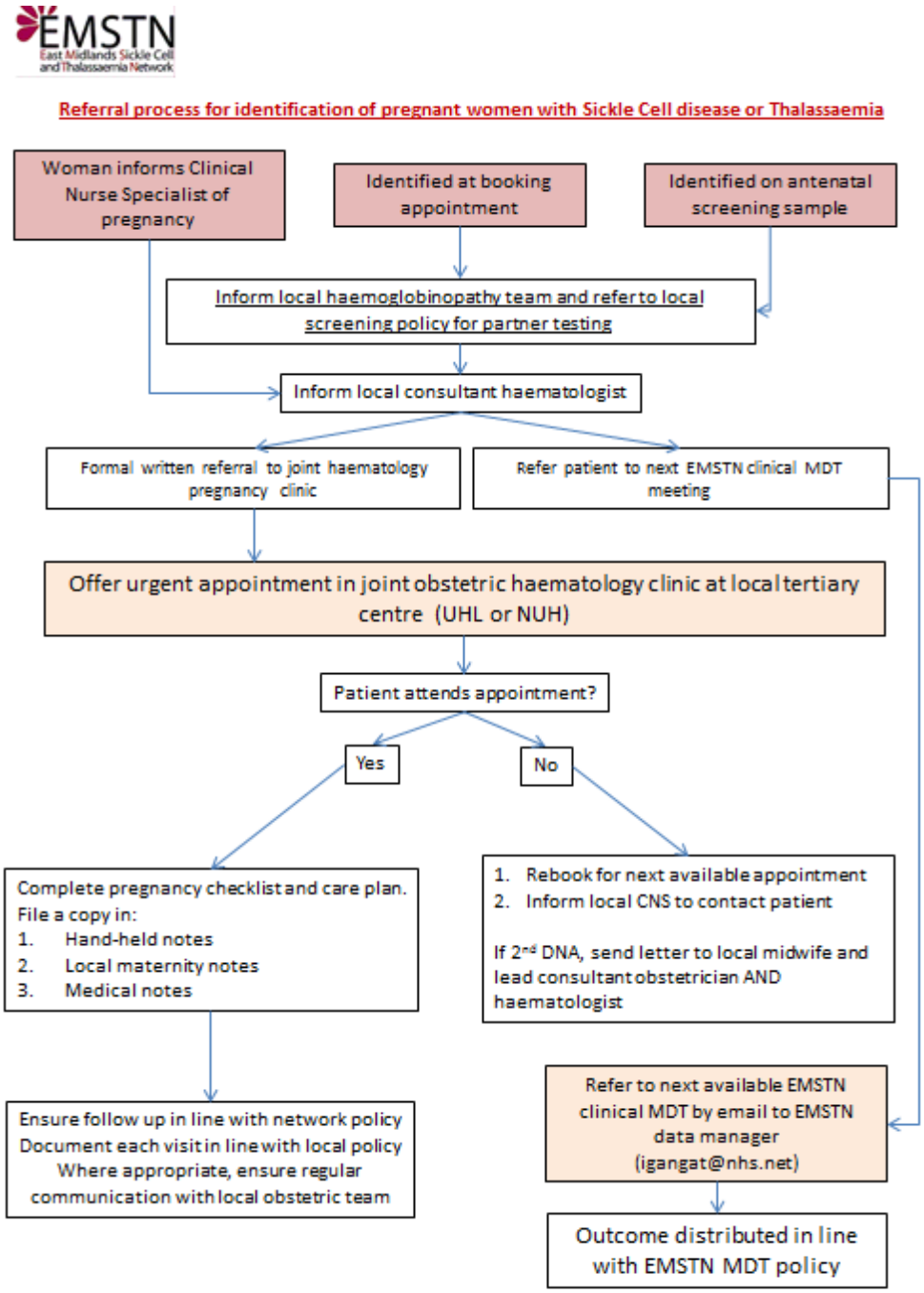
Blood group: **Red cell antibodies**: Y / N

Partner screening result:

Significant history:

If presenting in pregnancy, management plan should be implemented within 30 minutes If <24/40, admit under haematology team If ≥24/40, admit to labour ward		**Ensure both Haematology and Obstetric team informed**
1.	Initial Assessment	Document full set of observations (to include oxygen saturations on air) Keep warm to reduce risk of worsening of crisis
2.	Analgesia	Document pain score on admission and offer appropriate analgesia • Diclofenac 50mg STAT (if 12-28/40 gestation) then TDS for maximum of 2 days • Regular paracetamol (IV/PO depending on IV access) If moderate/severe pain: • Offer morphine sulphate SC/IV • If booking weight <50kg: Give 5mg STAT • If booking weight ≥50kg: Give 10mg STAT
3.	Fluids	500mls 4hrly (fluids may need to be warmed through blood warmer) Monitor regular fluid balance
4.	Oxygen	If oxygen saturation < 96% on air, give oxygen via mask at 5 l/min and ensure medical review (obstetric or haematology doctor)
5.	Heparin	Give LMWH prophylaxis in line with local policy during inpatient stay (unless contraindicated)
6.	Investigations	Blood tests: FBC, Retics, LDH, G&S, U&E, LFT Urine: Dipstick +/- PCR if proteinuria Microbiology: MSU, throat swab, blood cultures if suspected infection Imaging: Not for routine CXR – only perform if ? chest syndrome or ongoing concerns regarding infection
7.	Re-assessment	• Reassess pain at 30 minutes and repeat analgesia dose if no resolution; Ask regarding pain with each set of observations. • Repeat observations initially every 15-30 minutes to assess for opiate toxicity; if stable, perform in line with UHL policy • Assess for features of infection and if present, treat in line with local sepsis pathway
8.	Transfusion	**Unless clinical emergency, all transfusions should be discussed with the haematology team** If required, all blood should be HbS negative, Rh and Kell matched, CMV negative
9.	Ongoing medication	Ensure regular analgesia prescribed, to include PRN SC/IV/PO morphine up to 2 hourly, consider PCA if poor pain control. Continue regular folic acid 5mg OD and penicillin V 250mg BD (unless treatment antibiotics commenced). Screen for infection, consider IV antibiotics if febrile or features of sepsis
10.	Monitoring	**Concerns related to the following require escalation to the haematology on call team and early involvement of ITU. Patient may require emergency exchange transfusion.** • Opiate toxicity (low RR, reduced conscious level) • Chest syndrome (hypoxia with shadowing on CXR) • Hepatic/Splenic sequestration (often rapid fall in Hb with circulatory collapse) • Features of pre-eclampsia
11.	Obstetric Considerations	Consider need for CTG monitoring and escalate to obstetric team if concerns, particularly regarding reduced foetal movements. If admitted to labour ward, ensure obstetric anaesthetists aware of admission

Appendix 3: Referral process following identification of pregnancy



Appendix 4: Management plan for sickle cell crisis in pregnancy Reviewed 2022

Patient details (insert addressograph)

Patient diagnosis:

Baseline results: Hb Retics Sats on air

Blood group: **Red cell antibodies**: Y / N

Partner screening result:

Significant history:

<u>If presenting in pregnancy, management plan should be implemented within 30 minutes</u> If <23/40, admit under haematology team If ≥23/40, admit to labour ward **<u>Ensure both Haematology and Obstetric team informed</u>**		
1.	Initial Assessment	Document full set of observations (to include oxygen saturations on air) Keep warm to reduce risk of worsening of crisis
2.	Analgesia	Document pain score on admission and offer appropriate analgesia • Diclofenac 50mg STAT (if 12-30/40 gestation) then TDS for maximum of 2 days • Regular paracetamol (IV/PO depending on IV access) If moderate/severe pain: • Offer morphine sulphate SC/IV • If booking weight <50kg: Give 5mg STAT • If booking weight ≥50kg: Give 10mg STAT
3.	Fluids	500mls 4hrly (fluids may need to be warmed through blood warmer) Monitor regular fluid balance
4.	Oxygen	If oxygen saturation < 96% on air, give oxygen via mask at 5l/min and ensure medical review (obstetric or haematology doctor)
5.	Heparin	Give LMWH prophylaxis in line with local policy during inpatient stay (unless contraindicated)
6.	Investigations	Blood tests: FBC, Retics, LDH, G&S, U&E, LFT Urine: Dipstick +/- PCR if proteinuria Microbiology: MSU, throat swab, blood cultures if suspected infection Imaging: Not for routine CXR – only perform if ? chest syndrome or ongoing concerns regarding infection
7.	Re-assessment	• Reassess pain at 30 minutes and repeat analgesia dose if no resolution; Ask regarding pain with each set of observations. • Repeat observations initially every 15-30 minutes to assess for opiate toxicity; if stable, perform in line with UHL policy • Assess for features of infection and if present, treat in line with local sepsis pathway
8.	Transfusion	**Unless clinical emergency, all transfusions should be discussed with the haematology team** If required, all blood should be HbS negative, Rh and Kell matched, CMV negative
9.	Ongoing medication	Ensure regular analgesia prescribed, to include PRN SC/IV/PO morphine up to 2 hourly, consider PCA if poor pain control. Continue regular folic acid 5mg OD and penicillin V 250mg BD (unless treatment antibiotics commenced). Screen for infection, consider IV antibiotics if febrile or features of sepsis

10.	Monitoring	**Concerns related to the following require escalation to the haematology on call team and early involvement of ITU. Patient may require emergency exchange transfusion.** • Opiate toxicity (low RR, reduced conscious level) • Chest syndrome (hypoxia with shadowing on CXR) • Hepatic/Splenic sequestration (often rapid fall in Hb with circulatory collapse) • Features of pre-eclampsia
11.	Obstetric Considerations	Consider need for CTG monitoring and escalate to obstetric team if concerns, particularly regarding reduced foetal movements. If admitted to labour ward, ensure obstetric anaesthetists aware of admission

Intrapartum care plan for women with Sickle Cell Disease

Patient details (insert addressograph)	**Consultant Haematologist:** **Consultant Obstetrician:** **Date plan made:**
	Patient Diagnosis: **Booking Weight:** **EDD:** **Parity:**

Known Sickle Cell related Complications:	
Transfusion History:	Known Red cell antibodies? Yes / No
Antenatal events:	
Pain Management Plan:	Avoid Pethidine (lowers seizure threshold)
Plan for delivery:	Advised to omit LMWH if signs of labour or on day of planned delivery

Intrapartum Management:

Obtain IV access and send bloods for FBC/G&S; May require anaesthetic input if challenging

Inform haematology obstetric team of admission

Continuous foetal monitoring recommended

Keep warm and well hydrated; Monitor fluid balance and avoid fluid overload

Avoid prolonged labour (>12 hours) with early plan for escalation if poor progress

Monitor oxygen saturations and maintain >95%; Escalate to medical team if increasing oxygen requirements

Low threshold for antibiotics if signs of sepsis

Manage any sickle cell crisis as per antepartum plan; Ensure adequate analgesia and monitor response

Other:

Postpartum Management (Mother):

Keep warm, well hydrated and maintain sats >95%; Monitor fluid balance and avoid fluid overload

Encourage early mobilisation and restart LMWH prophylaxis 6-8 hours post-delivery (unless contraindicated)

Monitor for signs of crisis (higher risk if GA performed); Low threshold for antibiotics if signs of sepsis

Other:

Postpartum Management (Baby):

Monitor for signs of sedation, breathing difficulties, constipation, difficulty feeding and weight gain

Follow Neonatal plan if high risk for HDFN

If high risk for SCD, early testing should be offered

Discharge Plan:

Continue Thromboprophylaxis (Dose:……) for 6 weeks

Inform Haemoglobinopathy team on discharge (extension 16081) so that appropriate follow up can be made.

Completed by: **Signature:**	**Date:**