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Trust Guideline for the Screening and Management of Haemoglobinopathies in pregnancy

For use in:	Community, antenatal clinic and delivery suite
By:	Doctors and midwives
For:	Pregnant Women affected by Haemoglobinopathies
Division responsible for document:	Women and Children's Services
Key words:	Thalassaemia, sickle cell, screening, management
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Assessed and approved by the:	Maternity Guidelines Committee & NMCP If approved by committee or Governance Lead Chair's Action; tick here
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To be reviewed by:	Alison Evans, Daisy Nirmal
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Compliance links: *(is there any NICE related to guidance)*	None
If Yes - does the strategy/policy deviate from the recommendations of NICE? If so, why?	No

This guideline has been approved by the Maternity Guidelines Committee as an aid to the diagnosis and management of relevant patients and clinical circumstances. Not every patient or situation fits neatly into a standard guideline scenario and the guideline must be interpreted and applied in practice in the light of prevailing clinical circumstances, the diagnostic and treatment options available and the professional judgement, knowledge and expertise of relevant clinicians. It is advised that the rationale for any departure from relevant guidance should be documented in the patient's case notes.

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Version and Document Control:

Version Number	Date of Update	Change Description	Author
5	21/03/2022	1b renewed	Alison Evans, Daisy Nirmal
6	20/09/2022	1B flowchart has been updated	Alison Evans, Daisy Nirmal

This is a Controlled Document

Printed copies of this document may not be up to date. Please check the hospital intranet for the latest version and destroy all previous versions.

Quick Reference

1a – Antenatal SCT Pathway

1b – SC&T Linked Pathway

Objective

To ensure adequate support and care in pregnancy and labour for women with haemoglobinopathies.

Rationale

Sickle cell disease and thalassaemia major are serious, inherited blood disorders. They affect haemoglobin and its oxygen carrying capacity. Individuals who have one of these conditions need treatment and lifelong care

People who are carriers are healthy and unaware of their status unless they have a specific blood test

'Carrier' women and couples 'at risk' of having a baby with a major haemoglobin disorder need information, advice and counselling to make choices for the pregnancy. This includes the decision to have prenatal diagnosis and to take further action if they choose to.

Antenatal Screening at NNUH

All pregnant women should be offered screening for sickle cell and thalassaemia, ideally by 10+0 weeks of pregnancy. This is to allow women to obtain all the necessary information in time to make decisions about not continuing the pregnancy if wished.

NNUH is considered a low prevalence Trust by PHE guidelines. This means that all women have a full blood count as an initial screen, the laboratory then use the Family Origin Questionnaire (FOQ) to instruct on further testing. Where the woman or her partner falls into a high risk group on the FOQ, further variant testing is performed. Where a woman is identified as a carrier of a haemoglobinopathy, the laboratory will contact the Antenatal & Newborn Screening (ANS) team who will inform the woman of her result and offer partner testing. A failsafe process is in place between the laboratory and ANS team to ensure all women who are identified as carriers receive appropriate follow up and care.

Where a woman is identified as a carrier of sickle cell trait, an alert should be put on her medical and maternity notes as in some situations a partial crisis can be induced, such as use of certain anaesthetics.

Where a woman is identified as having a major condition ie. Sickle cell disease or beta thalassaemia major, she must be under the care of an Obstetrician leading on haemoglobinopathies in conjunction with a Consultant Haematologist due to the associated complications in pregnancy.

Where a couple are identified as both carriers of significant haemoglobinopathies they should be counselled by an appropriately trained professional e.g. Specialist Midwife, Consultant Haematologist, Consultant Obstetrician with special interest in Haemoglobinopathies. The couple should be informed they have a 1 in 4 risk of having an affected baby in each pregnancy. SCT programme information leaflets

should be used. Where the couple request prenatal diagnosis this should be offered ideally by 12+0 weeks of pregnancy. If the couple decline prenatal diagnosis, capillary testing at birth can be offered rather than waiting for newborn blood spot screening (see Linked pathway 1b) If a partner is unavailable for testing the woman should be counselled by an appropriately trained professional but capillary testing after birth would not be offered – newborn bloodspot screening is the recommended test where there is not a known 1 in 4 risk.

Screening Safety Incidents

Due to the nature and characteristics of screening tests, safety incidents within screening programmes require special attention and management. Where an incident occurs along any of the UKNSC screening pathways the ANSM should be informed and the UKNSC document “Managing Safety Incidents in NHS Screening Programmes: August 2017” referred to.

General Overview

Sickle Cell Disease in Pregnancy

- Increased incidence of
    - Perinatal mortality
    - Premature labour
    - Fetal growth restriction
    - Miscarriage
    - Early onset preeclampsia and pregnancy induced hypertension
    - Placental abruption
    - Fetal distress and Caesarean section
    - Thromboembolism
    - Maternal morbidity and mortality
    - Acute painful crises during pregnancy
    - Hospital admission

- There is also an increased risk of infection particularly of the urinary tract, pneumonia and puerperal sepsis. (Hyposplenism is common in these women and encapsulated organisms can cause overwhelming sepsis.)

Management of sickle cell disease in pregnancy

(See www.sicklecellsociety.org)
(See www.rcog.org.uk)

- Antenatal care should be provided by a multidisciplinary team including an obstetrician with maternal medicine involvement, midwife and haematologist
- End organ damage screen
- Avoidance of precipitating factors of sickle cell crises
- Recommend influenza vaccination
- Folic acid (5 mg/day) and penicillin prophylaxis (penicillin V 250 mg bd) should be given to all women.
- Consideration to prophylactic aspirin
- LMWH antenatally
- BP and urinalysis at every visit with monthly MSU
- Monthly assessment of Hb.
- Serial ultrasound assessment of fetal growth from 24 weeks
- Anaesthetic assessment in 3rd trimester
- Recommend induction of labour or caesarean section between 38-40 weeks gestation

Sickle cell crisis

This should be managed aggressively. This involves admission, adequate pain relief with opiates, adequate rehydration and early use of antibiotics if infection is suspected. The patient should be kept warm and well oxygenated. Arterial blood gases and oxygen saturation monitoring are mandatory.

Intrapartum avoidance of dehydration, hypoxia, sepsis and acidosis is important. Epidural analgesia is safe and advisable. Caesarean section should be performed for obstetric indications only. Liaise very closely with obstetric consultant on call, consultant haematologist and consultant anaesthetist.

The role of transfusion and exchange transfusion is controversial and should not be undertaken without discussion with a consultant haematologist.

Management of thalassaemia in pregnancy

- Women with alpha or Beta thalassaemia trait need iron and folate supplements throughout pregnancy.
- Blood transfusion may be required

Pregnancy in women with Beta thalassaemia major is very rare. Advice from a unit with particular expertise in the care of such women may be necessary. Features of iron overload due to repeated transfusions may result in hepatic, endocrine (diabetes, hypothyroidism) cardiac (left ventricular dysfunction) dysfunction and bone deformities due to bone expansion due to the drive to increased erythropoiesis.

Clinical audit standards

The directorate is committed to the regular audit of guidelines within available resources.

Summary of development and consultation process undertaken before registration and dissemination

The authors listed above drafted this guideline with the help of Dr. Hamish Lyall, Consultant Hematologist. During its development it has been circulated for comment.

This version has been endorsed by the Obstetric Clinical Guideline Committee.

Distribution list/ dissemination method

This guideline has been ratified by the O&G clinical guideline committee and has been disseminated via the hospital intranet to all members of obstetric staff.

References/ source documents

1. PHE Sickle cell and thalassaemia Screening Handbook 2018
2. PHE Sickle cell and thalassaemia handbook for antenatal laboratories Oct 2017
3. PHE Sickle cell and thalassaemia screening standards March 2019
4. Managing Safety Incidents in NHS Screening: August 2017
5. RCOG Green Top Guideline No. 61 – Management of Sickle Cell Disease in Pregnancy
6. RCOG Green Top Guideline No. 66 – Management of Beta Thalassaemia in Pregnancy