Ashford and St. Peter's Hospitals NHS
NHS Foundation Trust

WOMEN'S HEALTH AND PAEDIATRICS
MATERNITY UNIT

SICKLE CELL DISEASE - GUIDELINES FOR THE ACUTE MANAGEMENT OF PREGNANT WOMEN

Amendments			
Date	Page(s)	Comments	Approved by
Nov 2014		Complete document review	Women's Health Guidelines Group
March 2018		Complete document review – no changes	Women's Health Guidelines Group

Compiled by: Sandra Newbold, Consultant Obstetrician

In Consultation with: Obstetric and Anaesthetic Consultants and Senior Midwives

Ratified by: Women's Health Guidelines Group

Date Ratified: November 2014

Date Issued: November 2014

Next Review Date: March 2021

Target Audience: Staff working within maternity services

Impact Assessment Carried Out By: Women's Health Guidelines Group

Comments on this document to: Women's Health Guidelines Group

All women with sickle cell disease will be aware of their diagnosis, and which type of sickle cell disease they have. Their pregnancies will be jointly managed by an obstetrician and a haematologist and a management plan should be written in their obstetric notes.

However, they may be unexpectedly admitted with a crisis, dehydration secondary to vomiting, in preterm labour or with an intrauterine death; under these circumstances all management decisions will have to be made on the labour ward.

Problems occurring in women with sickle cell disease:

Present prior to pregnancy - pelvic arthropathy and/or avascular necrosis of the femoral head
- chronic renal failure
- chronic anaemia

45% fetal loss rate
- spontaneous abortion/IUD
- IUGR
- preterm delivery
- placental abruption

Maternal complications
- urinary tract infections
- pulmonary infections, especially pneumococcal pneumonia
- salmonella
- puerperal endometritis
- increased risk of pre-eclampsia.

The maternal mortality may be up to 3.3% (Howard et al, 1995) mainly due to cardiac failure, severe anaemia and thromboembolic complications.

Sickle cell crisis can be precipitated by infection, dehydration, over exertion and exposure to extreme temperatures (RCOG 2011). Pregnant women with sickle cell disease are advised to present to the labour ward if they have any symptoms of concern.

Sickle cell crises most commonly present with acute bony pain but can also present with abdominal pain. The management is essentially supportive with analgesia, rehydration, oxygenation and treatment of any precipitating factor such as infection. Less common, but much more serious, is a chest crisis. This is a rapidly progressive combination of pulmonary infection and infarction which carries a high mortality and requires urgent exchange transfusion and often ventilatory support. Progressing chest symptoms and signs must be taken seriously. Rarely aplastic crisis (where the bone marrow temporarily switches off) and sequestration crisis (which may be associated with hepatosplenomegaly) occur. Sickle crises may occur in patients who are S/C or S/beta thalassaemia double heterozygotes as well as in SS homozygotes.

All patients with sickle cell disease should be considered to be functionally hyposplenic and are therefore at particular risk from encapsulated organisms such as pneumococcus, haemophilus influenzae B and unusual organisms such as Salmonella sp.

If a woman with sickle cell disease is admitted to the labour ward Dr Bernard, or the on-call haematology consultant, and the labour ward or on-call obstetric consultant **MUST** be informed and involved in management decisions. The woman should be reviewed by a consultant obstetrician and consultant anaesthetist or haematologist within a few hours of admission.

MANAGEMENT OF A SICKLE CELL CRISIS

On presentation the woman should be assessed rapidly for medical complications requiring intervention such as ACS(acute chest syndrome) sepsis or dehydration. History should ascertain if this is typical sickle pain or not, and if there are precipitating factors. Examination should focus on the site of pain, any atypical features of the pain and any precipitating factors, in particular whether there are any signs of infection.

Investigations
- FBC, film and reticulocyte count
- G & S (Cross match 2 units of blood if has antibodies)
- urea, electrolytes and creatinine
- LFT including bilirubin and LDH
- Infection screen (blood cultures, MSU, HVS)
- CXR if clinically indicated.

AVOID USE OF TOURNIQUETS IF POSSIBLE

Treatment:

1. Analgesia: depends on severity of pain. Regular paracetamol should be prescribed. Strong opioids should be used if pain is severe. Consider morphine or fentanyl PCA. Pethidine should not be used because of the associated risk of seizures (RCOG 2011) NSAIDS are very effective in bony crises but should only be used 12 – 30 weeks (UKTIS 2014). Initial analgesia should be given within 30 minutes of arriving on the labour ward, and should be effective within 1 hour (RCOG 2011). Pain relief is often difficult and early discussion with the consultant anaesthetist or pain team is recommended.
2. Rehydration: intravenous fluids will usually be required. The usual regime will be Plasmalyte 1 litre 4 hourly initially, oral fluids can also be taken. Careful recording of fluid balance is vital, even if does not require intravenous fluids. Urea, creatinine and electrolytes must be checked 12 hourly. Potassium replacement depends upon these results.
3. Oxygen by face mask at 5 litres per minute, continuous 02 saturation monitoring is required and should maintain 02 saturation > 95% (RCOG 2011).
4. Intravenous antibiotics if infection suspected, usually Co-Amoxiclav, but appropriate antibiotic treatment should be discussed with a microbiologist. Ideally all sickle cell patients should take long term prophylactic penicillin or erythromycin, do not forget to restart the prophylactic antibiotics once the therapeutic course is finished.
5. Transfusion (replacement, exchange or partial exchange) if considered appropriate by the haematologist.
6. Folic acid 5mg daily (should be taking long term).

Sickle cell crisis is not an indication for delivery. Any delivery decision should be made purely on obstetric grounds. If at all possible delivery should be postponed until after the crisis has resolved.

MANAGEMENT OF LABOUR

These labours should be managed in the usual way but with careful attention to fluid balance, pain control and avoidance of infection. When a woman with sickle cell disease presents in labour:

1.Inform the labour ward or on-call obstetric consultant and either Dr Bernard or the on-call haematology consultant of admission.

2. Inform anaesthetic registrar that the woman is in labour. He/she should review the notes and see the woman early in labour.

3. Set up intravenous infusion - 4 litres Plasmalyte per 24 hours

4. Full blood count and G & S. (Cross match 2 units if the antenatal screen shows antibodies)
AVOID USE OF TOURNIQUETS IF POSSIBLE.

5. Keep careful record of fluid balance.

6. DVT prophylaxis - TED stockings
- s/c enoxaparin 40mg daily (usually starting after delivery)

7. Ensure adequate analgesia:
Entonox
TENS
Regular paracetamol
Epidural (provided > 12 hours since last Enoxaparin injection)
Morphine or Fentanyl PCA.

Pethidine should not be used because of the associated risk of seizures (RCOG 2011).

8. Antibiotics - to be started after delivery, or if catheterised during labour. In most cases Co-Amoxiclav (oral or if necessary intravenous) for 3 days is appropriate. If penicillin allergic use a cephalosporin and metronidazole. Prophylactic antibiotics do not need to be discussed with a microbiologist, but if there is any concern about infection this discussion must occur, and should be documented in the woman's notes.

Ideally all sickle cell patients should take long term prophylactic penicillin or erythromycin, do not forget to restart the prophylactic antibiotics once the therapeutic course is finished.

Postpartum:

1. Remember to inform paediatricians that a woman with sickle cell disease has delivered so that appropriate follow up of the baby can be arranged.

2. Continue intravenous fluids (as above) for 48 hours. She may still eat and drink, but decrease the rate of IV fluids if good oral intake.

3. Daily consultant review.
4. Keep careful record of fluid balance.
5. Antibiotics for 3 days (as above).
6. Post natal anticoagulation: dose depends on BMI. Give 7 days prophylaxis post vaginal delivery, 6 weeks prophylaxis after Caesarean section (RCOG 2011)

SICKLE CELL TRAIT

These women do not suffer any particular complications in pregnancy. They will be anaemic, iron supplements are only appropriate if the ferritin level is low.

Inform paediatricians that a woman with sickle cell trait has delivered so that appropriate follow up of the baby can be arranged.

Monitoring:

Compliance with this guideline will be monitored by reviewing all cases. Where monitoring has identified deficiencies, recommendations and an action plan will be developed.

References:

Howard RJ, Tuck SM, Pearson TC. Pregnancy in sickle cell disease in the UK: results of a multicentre survey of the effectof prophylactic blood transfusion on maternal and fetal outcome. *Br J Obstet Gynaecol* 1995;102;947–51.

Royal College of Obstetricians and Gynaecologists (2011). Management of Sickle Cell Disease in Pregnancy. RCOG; London. Greentop Guideline No 61.

UK Teratology Information Service (August 2014). Use of non-steroidal anti-inflammatory drugs (NSAIDs) in pregnancy. UKTIS 2014

EQUALITY IMPACT ASSESSMENT TOOL

Name: SICKLE CELL DISEASE - GUIDELINES FOR THE ACUTE MANAGEMENT OF PREGNANT WOMEN

Policy/Service: Women's Health and Paediatrics

Background • Description of the aims of the policy • Context in which the policy operates • Who was involved in the Equality Impact Assessment
To ensure consistent high standard of evidence base care Women's Health Guideline Group
Methodology • A brief account of how the likely effects of the policy was assessed (to include race and ethnic origin, disability, gender, culture, religion or belief, sexual orientation, age) • The data sources and any other information used • The consultation that was carried out (who, why and how?)
Policy widely circulated for comments within the Multidisciplinary Maternity Team.
Key Findings • Describe the results of the assessment • Identify if there is adverse or a potentially adverse impacts for any equalities groups
Accepted and understand the relevance of high standards of evidence based practice. Principles of equality have been adhered to.
Conclusion • Provide a summary of the overall conclusions
Improvement and consistency of maternity care provision
Recommendations • State recommended changes to the proposed policy as a result of the impact assessment • Where it has not been possible to amend the policy, provide the detail of any actions that have been identified • Describe the plans for reviewing the assessment
none

Guidance on Equalities Groups

Race and Ethnic origin (includes gypsies and travellers) (consider communication, access to information on services and employment, and ease of access to services and employment)	**Religion or belief** (include dress, individual care needs, family relationships, dietary requirements and spiritual needs for consideration)
Disability (consider communication issues, access to employment and services, whether individual care needs are being met and whether the policy promotes the involvement of disabled people)	**Sexual orientation including lesbian, gay and bisexual people** (consider whether the policy/service promotes a culture of openness and takes account of individual needs
Gender (consider care needs and employment issues, identify and remove or justify terms which are gender specific)	**Age** (consider any barriers to accessing services or employment, identify and remove or justify terms which could be ageist, for example, using titles of senior or junior)
Culture (consider dietary requirements, family relationships and individual care needs)	**Social class** (consider ability to access services and information, for example, is information provided in plain English?)

If further assessment is required please see the Integrated Single Equality Scheme.

For advice in respect of answering the above questions, please contact, HR Manager, on extension 2552.

PROFORMA FOR RATIFICATION OF POLICIES AND GUIDELINES BY RATIFYING COMMITTEE

Policy/Guidelines Name: Policy	**SICKLE CELL DISEASE - GUIDELINES FOR THE ACUTE MANAGEMENT OF PREGNANT WOMEN**
Name of Person completing form:	Dianne Casey
Date:	Nov 2014

Author(s)	Sandra Newbold
Name of author or sponsor to attend ratifying committee when policy/guideline is discussed	Dianne Casey
Date of final draft	Nov 2014
Has this policy/guideline been thoroughly proof-read to check for errors in spelling, typing, grammar and consistency?	Yes
By whom:	Women's Health Guidelines Group
Is this a new or revised policy/guideline?	revised

Describe the development process used to generate this policy/guideline.
Women's Health Guidelines Group, Labour Ward Forum, Obs & Gynae Consultants, Supervisors of Midwives
Who is the policy/guideline primarily for?
Health Professionals working within the maternity service
Is this policy/guideline relevant across the Trust or in limited areas?
Maternity Services
How will the information be disseminated and how will you ensure that relevant staff are aware of this policy/guideline?
Intranet, newsletters,
Describe the process by which adherence to this policy/guideline will be monitored.
See *monitoring section of policy*
Is there a NICE or other national guideline relevant to this topic? If so, which one and how does it relate to this policy/guideline?
See *reference section of policy*
What (other) information sources have been used to produce this policy/guideline?
See *reference section of policy*
Has the policy/guideline been impact assessed with regard to disability, race, gender, age, religion, sexual orientation?
No impact
Other than the authors, which other groups or individuals have been given a draft for comment?
All obstetric Consultants, Women's Health Guidelines Group, SOM's
Which groups or individuals submitted written or verbal comments on earlier drafts?
Any comments received considered by Women's Health Guidelines Group
Who considered those comments and to what extent have they been incorporated into the final draft?
All comments considered
Have financial implications been considered?
Yes