Brighton and Sussex University Hospitals NHS Trust

Care of Women with Haemoglobinopathies

Maternity Protocol: MP036
Date agreed: January 2016

As discussed at previous audit and safety meetings as part of the review of guidelines following the review of women's services governance Julie Wade was attached to the governance team to undertake some guideline reviews. At the back of the guidelines Julie has attached a clear summary of her review. These guidelines will be circulated on a daily basis for your comments.

Whilst is acknowledged that in an ideal world only a few guidelines would be sent out for comments however unfortunately as we need to get all our guidelines in date and be safe and fit for purpose we do not have that luxury. Further it also recognised that Julie is a midwife and some of these guidelines are obstetric focused. We are very grateful for everyone's support in taking our guidelines forward.

Guideline Reviewer:	Julie Wade
Manager responsible:	Heather Brown
Version:	2.1
Approval Committee:	Women's Safety and Quality Committee
Date agreed:	January 2016
Review date:	January 2019
Cross reference:	MP001 Antenatal booking and risk assessment MP035 Care of women in Labour

Contents

Key Principles

A protocol is a set of measurable, objective standards to determine a course of action. Professional judgement may be use in the application of a protocol.

Scope

♦ This protocol applies to:
- Childbearing women with Haemoglobinopathies

Responsibilites

- Midwives & Obstetricians:
  - To access, read, understand and follow this guidance
  - To use their professional judgement in application of this protocol
- Management:
  - To ensure the protocol is reviewed as required in line with Trust and National recommendations
  - To ensure the protocol is accessible to all relevant staff

Objective Standards

1.0 Haemoglobinopathies in pregnancy

All pregnant women with haemoglobinopathies should be discussed with the antenatal screening co-ordinator midwife. They should be offered partner testing after a discussion about pre-natal diagnosis. This discussion should happen as soon as possible after the abnormal result and ideally within the first trimester.

1.1 Thalassaemia in pregnancy

Thalassaemia trait

- The thalassaemia syndromes are named and classified by the type of haemoglobin chain that is inadequately produced. Apart from mild, hypochromic, microcytic anaemia, both α- and β-thalassaemia traits do not present any hazard to the mother or the fetus. However, prenatal diagnosis may be important if the partner is similarly affected.

Thalassaemia major and intermedia

- Beta thalassaemia major and intermedia is associated with an increased risk to both mother and baby, such as cardiomyopathy in the mother due to iron overload and the increased risk of fetal growth restriction. Women with thalassemia major may also develop diabetes or hypothyroidism due to the increasing iron burden. A haematologist should be involved in the antenatal care of these women.

- Women should be seen by Maternal Medicine in the first trimester. A management plan regarding the frequency of their ongoing care should be made. 4 weekly fetal scans from 24 weeks gestation are recommended.

- All women with thalassaemia major should have a cardiac assessment at 28 weeks gestation and thereafter as appropriate.

- Thyroid function should be checked periodically.

- Transfusion should be considered for worsening maternal anaemia or evidence of fetal growth restriction.

- Women should be advised to take 5mg Folic acid daily. (RCOG 2014)

1.2 Sickle cell disease (not trait) in pregnancy

- Haemolytic and painful crisis is commonest in the third trimester. Crisis can be precipitated by stress, pain, dehydration, reduced oxygen tension and infection. Advising women to avoid the above where possible and to seek early medical advice (RCOG), as well as regular observations, urine testing, and infection screening are therefore essential. Intravenous infusion may be required to prevent dehydration. ***Always believe the level of pain shown.***

- Women may be at increased risk of pre-eclampsia and fetal growth restriction, therefore: women should have serial growth scans from 24 weeks, and be considered for low dose aspirin 75mg daily from 12 weeks gestation.

- A haematologist should be involved in the antenatal care of these women. (RCOG)

- Transfuse only if and when a woman develops a recurrent or severe SCD related complication.

- Transfuse those women with a history of severe sickle cell disease, or obstetric problems.

- Low molecular weight heparin should be started at 36 weeks gestation. (RCOG)

- Advise 5mg folic acid daily throughout pregnancy. (RCOG)

Note:

1. Careful and regular obstetric and haematological follow-up is essential
2. The Hb may fall to 6 g/dl, or less, at the end of the second trimester. If the mother is feeling well, there is no need to transfuse to correct the Hb.
3. If transfusion needed, use genotyped and leukocyte depleted blood
4. Pregnancy may be complicated by aseptic necrosis of the hip and worsening retinopathy

1.3 Management of antenatal sickle cell crisis.

- Admit to delivery suite
- Refer to obstetric and anesthetic team

- Take full infection screen (HVS, MSU, sputum, FBC and blood culture)
- Check FBC, U&E + LFT, quantitative electrophoresis
- Provide adequate analgesia
- Give oxygen by facemask
- Site IV cannula, rehydrate and transfuse if Hb<8.0g/dl
- Monitor continuously with CTG
- Use pulse oximetry

2.0 Management of haemoglobinopathies in labour

2.1 Thalassaemia (major and intermedia, not trait) in labour

- There is no contraindication to normal obstetric practice, or to epidural anaesthesia. Vaginal delivery whenever possible.
- On admission take blood for full blood count and group and save. If Hb is <100g/l, cross match 2 units blood.
- Continuous electronic fetal monitoring should be performed.
- Active management of the third stage of labour is recommended. (RCOG)

2.2 Sickle cell disease (not trait) in labour

- There is no contraindication to normal obstetric practice, or to epidural anaesthesia. Vaginal delivery whenever possible.
- Inform Obstetrician of admission.
- Take maternal blood for FBC, quantitative electrophoresis, and cross-matching.
- Continuous electronic fetal monitoring should be performed. (RCOG)
- Check each urine specimen for proteinuria as this may be a sign of impending crisis

- Administer prophylactic antibiotics.
- Encourage epidural rather than Pethidine for analgesia.
- Give oxygen by mask, continuing for 36 hours post-delivery. If GA, CPAP for at least 24 hours, preferably on ITU.
- Inform anaesthetist.
- Use pulse oximetry.
- Maintain hydration with IV fluids.
- Give Ranitidine 150mg orally 6 hourly in labour.
- Ensure Paediatric support is present at delivery.
- Take cord blood for FBC and electrophoresis.
- If caesarean section is necessary, oxygen is essential and partial exchange transfusion may be required. Ensure anaesthetist is aware of the problem.

3.0 Postnatal management of haemoglobinopathies

3.1 Venous thromboembolism risk

Women with these haemoglobinopathies should be considered high risk for venous thromboembolism and should have low-molecular-weight heparin (for 7 days after vaginal delivery, 6 weeks after caesarean section).

4.0 References

1. Aessopos A, Karabatsos F, Farmakis D, Katsantoni A, Hatziliami A, Youssef J, Karagiorga M. Pregnancy in patients with well-treated beta-thalassemia: outcome for mothers and newborn infants. Am J Obstet Gynecol. 1999; 180:360-5.

2. Jensen CE, Tuck SM, Wonke B Fertility in beta thalassaemia major: a report of 16 pregnancies, preconceptual evaluation and a review of the literature. Br J Obstet Gynaecol. 1995; 102:625-9

3. Royal College of Obstetricians and Gynaecologists (2011) *Green-top guideline no. 61. Management of sickle cell disease in pregnancy*. London. RCOG. www.rcog.org.uk

4. Royal College of Obstetricians and Gynaecologists (2014) *Green-top guideline no. 66. Management of beta thalassaemia in pregnancy.* London. RCOG. www.rcog.org.uk