Haemophilia Carriers – Management of Women in Pregnancy, Intrapartum and the Postnatal Period

University Hospitals of Leicester NHS Trust

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1. Introduction and who the guideline applies to:

The wide range of symptoms and effects from haemophilia make comparisons of the published literature challenging and limit the ability to provide detailed recommendations for specific aspects of care.

This guideline provides guidance for midwives, medical and support staff on the different management choices and the options available for women who are carriers of Haemophilia.

2. Management of Haemophilia in pregnancy

2.1 Pre-conception and Antenatal

- All women should be offered counselling as they are at risk of;
  (a) serious bleeding postpartum,
  (b) giving birth to an affected male
- Women should be offered pre-conception counselling either within the haematology clinic or the haematology obstetric clinic.
- The risks to the fetus and the women must be discussed.
- Screening and diagnostic testing should be discussed and a plan of pregnancy care mentioned. In carriers of severe haemophilia this should include offer of pre-implantation genetic diagnosis.

2.2 Investigations and monitoring

- Baseline factor levels (factor VIII/IX) should be checked pre-conceptually
- Baseline FBC levels should be checked pre-conception if not already done
- If baseline VIII/IX levels are <0.5 iu/ml it is important to ensure that the women are vaccinated against hepatitis A and B (if not already immune) prior to embarking on a pregnancy.
- General health should be optimised pre-conception including weight and correction of iron deficiency.

2.3 Prenatal testing

- All women should be offered prenatal diagnosis in pregnancy
- All women should be offered prenatal testing but women who do not wish prenatal diagnosis should be encouraged to have fetal sex determined to allow safe planning of pregnancy and delivery.
- Fetal sex can be determined by ffDNA from 9 weeks gestation.
- Carriers of severe haemophilia with a male fetus should be offered prenatal diagnosis by CVS at 11-14 weeks gestation.
- All haemophilia carriers should be offered a third trimester amniocentesis for diagnosis.
- Following the anomaly scan or prenatal testing results the women should be seen in the haematology obstetric clinic in order that a plan of care in pregnancy and delivery may be documented.

2.4 Monitoring during pregnancy

- Factor levels should be checked throughout pregnancy and prior to any invasive procedures.
- Levels should be checked at booking, 28 and 34 weeks and before invasive procedures (factor VIII levels usually rise in pregnancy but factor IX tends to remain constant).
- Aim for factor levels of >0.5 iu/ml to cover surgical and invasive procedures or spontaneous miscarriage. The haemostasis team will advise on haemostatic treatment required to achieve this.
- If factor levels are <0.5iu/ml the haemostasis team may give treatment with recombinant clotting concentrate or DDAVP (haemA) for potentially haemorrhagic events such as invasive diagnostic procedures, spontaneous abortion, TOP and labour. Pre and post levels should be checked as directed by haemostasis team to ensure therapeutic levels are maintained for a suitable time period depending on the procedure.
- DDAVP may be considered with discussion with haematology team. This requires a fluid restriction of 1 litre in 24 hours.
- Tranexamic acid should be considered in combination with haemostatic treatment in those with levels of <0.5 iu/ml and alone for those with levels of >0.5 iu/ml. Following miscarriage, this should continue until bleeding stops.
- There should be a documented plan for delivery completed by 34 weeks
- Mode of delivery should be agreed jointly between the woman and the MDT.
- For intended vaginal delivery, spontaneous labour is preferred, if no other obstetric concerns, to minimise the risk of intervention.
- The option of planned C-section should be discussed for the delivery of affected male babies, especially in those with severe haemophilia, and/or if fetal status is unknown. This should involve full assessment of advantage and disadvantages.
- If elective C-section is planned, this should be at 39 weeks gestation with a clear plan for spontaneous labour occurring beforehand.
- Pregnant women should be seen by anaesthetics antenatally.

Fetus

- A paediatric alert should be put in place and referral to neonates antenatally considered.
- Third trimester (35-36 weeks) amniocentesis should be considered if undiagnosed male fetus at risk of severe Haemophilia

2.5 Intrapartum

Blood tests that should be performed:

- FBC
- PT, APTT and fibrinogen
- Group and save
- Factor assay if this was <0.5 iu/ml at last check. Treatment should be given if levels are <0.5 iu/ml

If admitted in spontaneous labour, please inform haematology team on-call.

- Ensure minimal trauma to both mother and baby. Avoid fetal blood sampling, fetal scalp monitoring. Ventouse delivery, mild cavity forceps involving rotation of the head. External cephalic version should be avoided in potentially affected male babies.
- Active management of the 3rd stage of labour is recommended.
- If additional haemostatic treatment is required for delivery, this will be guided by the haemostasis team and should be given as close to delivery as possible.
- Tranexamic acid should be considered until lochia is minimal.
- Regional anaesthesia is known to be safe if the coagulation screen is normal and factor levels are >0.5 iu/ml (Letsky 1991,Kadir 1997) but levels must be checked prior to removal of catheter as they fall rapidly after birth.
- If maternal FVIII/IX levels <0.5 iu/ml avoid NSAIDs, and aspirin and IM injections. (Mannucci 2005)

2.6 Postpartum

- A cord sample should be sent for factor level in male neonates and ensure the results are known **BEFORE** the patient leaves hospital.
- Be aware that factor VIII and IX from the newborn do not always reflect the true baseline level. This may require retesting at 3-6 months of age.
- IM vitamin K should not be given to the baby until the results are known or give orally. Avoid other IM injections to the baby.
- Cranial ultrasound should be considered prior to discharge in all neonates with a history of moderate or severe haemophilia.
- The woman should be observed for postpartum haemorrhage as levels fall rapidly after delivery. Maintain levels >0.5 iu/ml for 3-4 days or longer if C-section has been performed.
- DDVAP may be used immediately postpartum.
- Any decision about thromboprophylaxis will be made by the obstetric haematology team on an individual patient basis.

3. Education and Training

None

4. Monitoring Compliance

None

5. Supporting References

RCOG guideline no 71 – management of inherited bleeding disorders in pregnancy.
Letsky EA Haemostasis and epidural anaesthesia. int j obst anaesthesia 1991:1:51-54
Kadir RA, Economides Dl, Braithwaite j, goldman e, Lee c the obstetric experience of carriers of haemophilia British J obs gyn 1997; 104:803-810

PM Mannucci Use of desmopressin (DDAVP) during early pregnancy in factor VIII– deficient women. Blood 2005; 105:3382

Pavord s, Hunt B The Obstetric Haematology Manual

6. Key Words

Haemophilia carriers in pregnancy Haematology / obstetric team

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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	
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Details of Changes made during review: **Format update only**	