Obstetric and Neonatal Management of Carriers of Haemophilia A and B – Summary Clinical Guideline

Reference No.: Obst/07:23/H2

In women who are known carriers of haemophilia the opportunity exists to manage pregnancy, labour and delivery and the early neonatal period in order to minimize the increased risk of bleeding in both the mother and the affected fetus/neonate. Successful management of both mother and baby depends on a multidisciplinary approach (obstetric, haematological, neonatal and midwifery)

Pre-pregnancy care

Females with a family history of haemophilia, known carriers or those with partners with haemophilia should be offered referral to the combined obstetric haematology service for pre-pregnancy counselling and referral for genetic counselling and screening for carrier status where appropriate. The identification of a molecular marker in the family is necessary if prenatal diagnosis is to be considered in pregnancy.

Pre-implantation genetic diagnosis (PIGD) by IVF can be utilized in haemophilia and may be considered by some couples in preference to prenatal diagnosis. Couples wishing to consider this option would need referral to a local fertility specialist and would require discussion with the Clinical Commissioning Group (CCG) regarding funding.

Pregnancy Management

Refer for early booking under combined obstetric haematology service +/- to Fetal Medicine Consultant if wish to consider first trimester prenatal diagnosis by CVS. The latter is only available to women in whom a molecular marker has been identified.

A plan for Labour/delivery should be discussed antenatally between the Consultant Obstetrician and the mother, taking into account obstetric issues, maternal factor levels and the known or potential haemophilia status of the fetus.

The women should be referred antenatally to the obstetric anaesthetic clinic to discuss pain relief options and anaesthesia.

Any plan of care to be documented in the maternal records.

A neonatal alert should be completed if the mother is carrying an affected or potentially affected male child.

Labour / delivery care

On admission check FBC, Clotting, G&S and Factor VIII/IX if < 50% at last check or if greater than 2 weeks since last.

Aim for vaginal delivery unless obstetric contraindication to this or antenatally determined plan recommends Caesarean Section. Spontaneous labour is preferable to

induced labour as the latter is likely to be longer and have an increased risk of need for operative delivery.

Avoid maternal IM analgesia if level< 40%. Pethidine may be administered subcutaneously if necessary, at the same dosage. The onset time may be slightly longer than for IM administration.

Intrapartum management decisions should be made in conjunction with a Consultant Obstetrician or other suitably experienced Obstetrician to minimise the risk of traumatic bleeding for both mother and baby. If the fetus is confirmed unaffected after prenatal diagnosis or is female, normal intrapartum care should be offered. If the fetus is a potentially affected male or of unknown sex the labour/delivery should be managed as if affected fetus and procedures that carry an increased risk of cranial bleeding should be avoided where possible.

- Principles of management are to achieve the least traumatic delivery possible utilising senior midwifery and medical staff
- Avoid FSE/ Fetal blood sampling
- Normal vaginal delivery should be conducted by an experienced midwife to reduce the risk of perineal trauma
- Ventouse, rotational and mid cavity forceps carry a significant increase in the risk of cranial bleeding and should be avoided unless delivery by C/S considered more traumatic.
- Low cavity forceps likely to be less traumatic than a full dilatation C/S but should only be undertaken by a senior obstetrician.
- Early recourse to Caesarean Section if significant delay in labour
- Active management of the third stage should be practised, physiological management is contraindicated
- Prompt repair of perineal trauma by suitably experienced operator
- There is little data to inform management of breech presentation but there would be concerns about the risk of ICH with both ECV and vaginal breech birth

Baby

If male infant, a cord sample should be taken in a citrate tube for coagulation screening and Factor VIII/IX assays after discussion with Haematology Consultant.
Where severe haemophilia A or B is suspected the diagnosis should be confirmed by factor assay within the first few hours after delivery.
If there is any uncertainty about contamination of the cord sample with maternal blood a venous sample should be obtained from the baby.

Avoid IM injections or heel stab until clotting factor results known. Give Vitamin K orally if any significant delay anticipated in obtaining result.