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Pregnancy and Delivery Management – a Nursing Perspective

Robyn Shoemark

Clinical Nurse Consultant

Haemophilia/Haematology

The Kids Factor Zone

The Children's Hospital at Westmead

Full disclosure

I am a paediatric nurse ☺

Nursing Role

- Important to know your patient and family
- Regular discussions
- Carrier status
  - Obligate carriers
  - Who should we test
  - When to test girls
  - How do we keep track of testing
- Counselling – preferably pre pregnancy

Planning a pregnancy?

- Do you have a haematologist?
- Do you know your carrier status?
- Do you know your baseline factor levels?
- Genetic counselling
  - Information
  - Allows for reproductive options
    - Natural conception
    - PGD

https://www.genetics.edu.au/SitePages/Genetic-Services.aspx

You are pregnant

- Congratulations
- Notify your haematologist and HTC
- Pre pregnancy factor levels
- Do you want the pregnancy tested
- Birthing plan
  - When
  - Where
  - Precautions

Notify HTC

- Sooner better than later
- Able to check carrier status if not previously done
- Discuss options
- Arrange referrals
- Make a plan

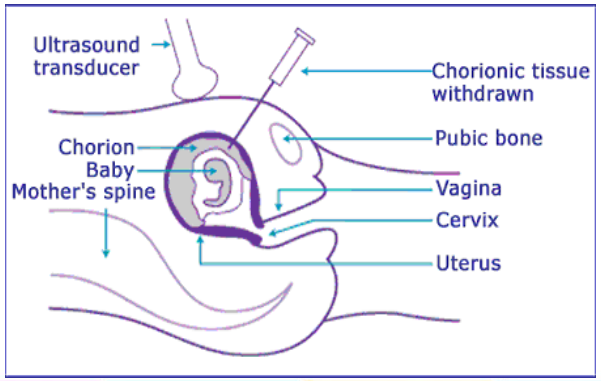
Your levels?

- Normal factor levels
  - Lower risk to mother
- Low factor levels
  - Higher risk to mother

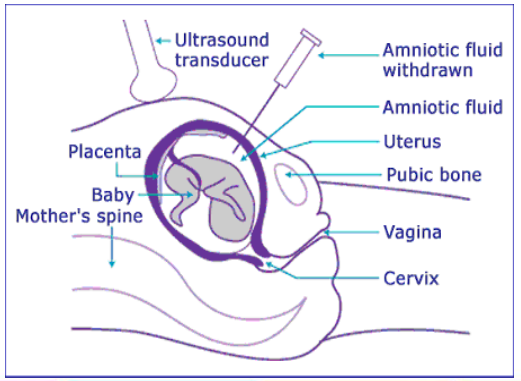
Testing

- No testing
- Cell-Free foetal DNA or NIPT (>9 weeks)
- Ultrasound (18-20 weeks)
- Chorionic villus sampling (11-14 weeks)
- Amniocentesis (15-19 weeks)

CVS

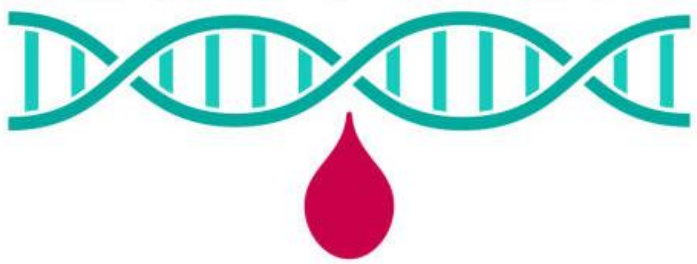


Amnio



Where?

- Private hospital
- Public hospital
- Hospital with a high-risk birthing unit
- Hospital with both high-risk unit & HTC
- Informed and educated decision



Information

Letters, genetic information +/- sex of baby and pre pregnancy factor levels

- Sent to high-risk birthing unit
- Sent to obstetrician
- Mother to have a copy

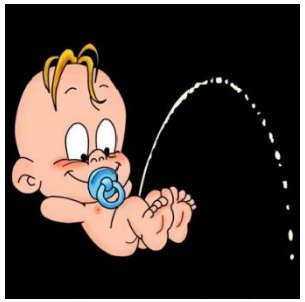
Mum plan

- Personalised plan to include
  - Regular monitoring of factor levels
  - Birth at a hospital that has a HTC and a haematologist
  - Pre and post birthing factor plan which is decided by the haematologist. This may include tranexamic acid, DDAVP or factor

With regard to her pregnancy as per the Australian consensus statement on management of inherited bleeding disorders in pregnancy:

- *___levels may or may not increase during pregnancy but can drop in the postpartum period; and will need to be monitored during her pregnancy and the immediate post-partum period.*
- *Her levels should also be checked before any invasive procedures and factor replacement therapy considered if her levels are below 50%.*
- *The mode of delivery should be determined by obstetric indication. However, vacuum extraction is contraindicated due to the high risk of intracranial and subgaleal haemorrhage. The use of forceps, foetal scalp blood sampling and scalp electrode should be avoided if possible.*
- *When ___ is admitted to hospital , please contact the haematology registrar.*
- *___ will have ___IU of factor with her, which she will bring to hospital (I have given her factor today and she will bring it to hospital at the time of delivery).*
- *At the start of the 2nd stage of labour, I would suggest that ___ will have factor ___IU*
- *Tranexamic acid 1g prior to delivery*
- *___ should also have factor ___IU for 3 days post-partum if vaginal delivery and for 5 days if a caesarean section is performed.*

Baby plan



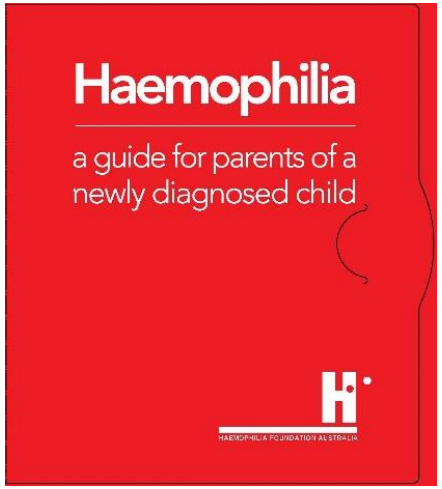
- **If sex known**
  - Female baby usually able to birth locally with plan to call Haematology for advice as needed
  - Male baby known to be affected, advised to birth at hospital with HTC
  - Male baby but not known if affected, advised to birth at hospital with HTC, assuming baby has bleeding disorder until proven otherwise
- **If sex unknown**
  - Assume to be an affected male and advised to birth at hospital with HTC

Our recommendations for any pregnancy at risk of a baby with haemophilia include the following:

1. *Delivery at a tertiary centre preferably one with a Haemophilia Treatment Centre attached (Westmead is ideal) where there is access to factor products and staff experienced in managing bleeding disorders.*
2. *Delivery: An atraumatic delivery as possible (ideally without instrumentation) is preferred. Vaginal delivery is acceptable provided it can be assessed as likely to be not too traumatic. Generally speaking, the need for a Caesarean Delivery should be decided on obstetric grounds.*
3. ***Ventouse extraction, rotational and mid-cavity forceps are associated with an increased risk of bleeding and should be avoided***
4. *Invasive monitoring procedures such as placement of intrapartum scalp electrodes and foetal scalp blood sampling should be avoided.*
5. *Ideally the paediatric team should be at the delivery to assess the baby for any signs of bleeding*
6. *Collect cord blood to test for APTT and factor VIII levels*
7. *IM injections of vitamin K and hepatitis B vaccine should not be given to the baby*
8. *Oral vitamin K should be given. Hepatitis B vaccination given subcutaneously in the first week of life - we will typically do that when we see the baby here at CHW.*
9. *The baby needs to be seen by the paediatric team promptly to assess for any potential bleeding (especially intracranial) with clinicians having a low threshold for performing a head ultrasound.*
10. *Contact the haematology team at the Children's hospital (9845 3304 (registrars)/ 9845 0839 (Haemophilia CNC)) once the baby is born and the Factor VIII level is known for further advice or earlier if there are concerns.*

After the birth

- Care for the Mum
  - if she has a bleeding disorder, follow haematologists plan
- Care for baby
  - Script for oral Vitamin K arranged
  - Hep B injection before day 7
  - Introduction to HTC if first baby
  - Information and education



Quote

Since finding out our baby was affected by Haemophilia, we knew straight away we would prefer to deliver at Westmead so we could be close to the Children's Hospital, if required. I made contact with Robyn the Haematology Nurse who organised a Doctor to write the referral and organise the transfer. The process was seamless, and all done within a couple of weeks.

The care I have received since transferring to Westmead has been fantastic. I have the same midwife and Doctor I see at every visit. I have the midwife's direct phone number and never wait in the waiting room. It's like VIP service!

It takes a village

Thank you