PATHWAY FOR PREGNANT BIRTHING PEOPLE WITH INHERITED BLEEDING DISORDERS (IBDs) OR THOSE WITH POTENTIALLY AFFECTED BABIES WITH BLEEDING DISORDERS

REFER AS SOON AS POSSIBLE IN PREGNANCY

Email: ohtc.secretaries@ouh.nhs.uk and refer to the Silverstar Unit silverstar.midwives@oxnet.nhs.uk

Please provide the following information if possible

- Name:
- Date of birth:
- NHS number:
- Address:
- Email/Telephone number of patient:
- Type of inherited bleeding disorder and severity: who is affected in the family?
- Baseline factor levels/FBC in pregnant person, Blood Group, RhD status
- Baseline factor levels/FBC in affected individuals with the bleeding disorder
- ISTH BAT score if available
- Causative genetic variant if known and inheritance of disorder/Family Number
- Treatment usually given for bleeding disorder if affected
- EDD –scans/LMP
- Does the patient wish to continue with the pregnancy?
  - If no: urgently email ohtc.reception@ouh.nhs.uk and arrange dating scan ASAP locally
- If the pregnancy is not viable or the patient is having a miscarriage: please urgently call the on call the Haemophilia Consultant via the John Radcliffe Hospital switchboard: 0300 304 7777
- Other comorbidities
- Complications in this and prior pregnancies including bleeding
- Height, Weight, Blood pressure:
- Any communication/safeguarding issues
- Local obstetric team contact details

Community midwives/GPs
Oxford Haemophilia and Thrombosis Centre (OHTC) Regional Haematology/Obstetric Teams
Silver Star/Paediatric Haemophilia Unit/OUH Teams
Patient self referral to OHTC

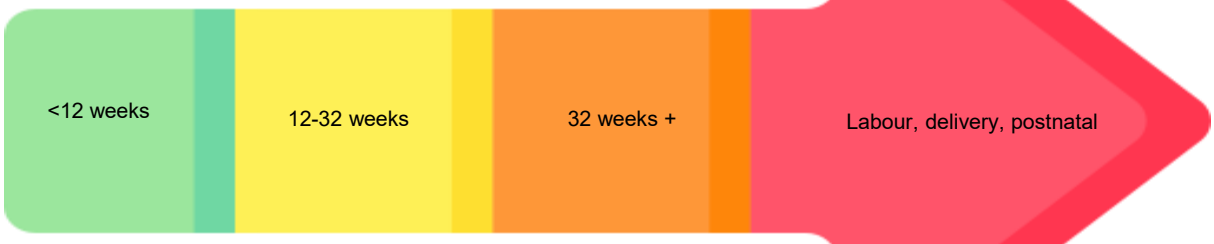
REFER AS SOON AS POSSIBLE TO OHTC
IF THE PATIENT HAS TO DELIVER IN OXFORD PLEASE ALSO REFER DIRECTLY TO SILVER STAR

Rationale for early referral

- Patient may need treatment for invasive tests or bleeds
- Patient may require time critical tests and subsequent safe management: e.g. fetal sexing, CVS, termination
- Patient's carrier status/bleeding disorder status might not be known: early referral allows for genetic testing and return of results and appropriate management/counselling
- Many women who are carriers or have IBDs are iron deficient and require early replacement to prevent maternal and fetal morbidity
- Referral may be needed to fetal medicine, maternal medicine (Silver Star), paediatric haemophilia as well: please copy in both teams

Rationale for calling haematology when the patient is admitted

- The patient may require factors/DDAVP and these might not be available on delivery suite
- The patient may require levels to be measured and the labs and BMSs need to liaise to turn on the analyzers (out of hours) but also collect blood samples and ensure these are not lost
- If there is a delay due to failure of progression between administration of factor/treatment and delivery: more levels, further dosing may be required before delivery/removal of epidural or spinal anaesthesia
- Both adult and paediatric teams can be primed and familiar with the plan if there is maternal or fetal haemorrhage so that timely and correct haemostatic treatment is given
- Postnatal haemostatic treatment may be required and we can advise on dosing and timing of that, dependent on delivery outcome and VTE thrombo-prophylaxis
- Haematology can arrange postnatal Ward round review so that there are no delays to discharge, including paediatric haemophilia review of affected babies.



- Clinic review of diagnosis/genetics
- Initial counselling about management in pregnancy, prenatal diagnostics, delivery plans
- Factor level check
- FBC, Iron profile check
- Blood group RH D confirmation
- Dating scan if termination considered urgently to confirm intrauterine pregnancy
- Free fetal DNA sexing (ffDNA) if termination considered (9 weeks by scan)
- Genetic testing if status unknown
- Referral to Silver Star if needed

If ffDNA confirms male fetus and termination being considered, refer for CVS

If CVS confirms affected male: arrange termination at maternal request

Ensure blood group RHD status and factor levels and haemostatic plan provided

- Usually seen after 20 week US scan to confirm fetal sex if ffDNA not done previously
- Clinic review of diagnosis/genetics
- Counselling about management in pregnancy, prenatal diagnostics, delivery plans
- Factor level check
- FBC, Iron profile check
- Blood group RH D confirmation
- Referral to Silver Star if needed
- Referral to Fetal Medicine for late amniocentesis after counselling if appropriate

- Late amniocentesis if requested
- Finalise obstetric haemostasis plan and send to patient and DGH teams, CHOX and complete maternity situational awareness alert
- Ensure factor/haemostatic medication available in blood banks and maternity units
- Factor level check
- FBC, Iron profile check: arrange IV iron if inadequate response to PO iron (ID without anaemia and IDA)
- Ensure supply of TXA
- Cord pack given with forms and consent taken for cord genetics if appropriate
- Book day 10 and week 6 follow up with OHTC

INFORM HAEMOPHILIA TEAM OF ADMISSION AS PER PLAN

- Call if excess bleeding, unexpected bleeding, protracted bleeding
- If baby might be affected by Haemophilia: await cranial US
- **DO NOT DISCHARGE BABY AND MOTHER until**

1. **Haemophilia team review**
2. **Cord levels/ peripheral blood levels/FBC back if taken and discussed**
3. **Paediatric haemophilia review for possible affected babies**
4. **Provision for further doses of PO vitamin K made if required**

- OHTC will call women day 10 postnatal and review week 6 postnatal: review lochia, iron profile, wound healing
- OHTC Consultant will chase cord blood genetics and contact mother with results
- Confirmed bleeding disorders on cord genetics patients will be registered by paediatrics when reviewed in clinic
- Possible carriers will be registered at 6 week check by adult OHTC team

- Acronyms:
- CHOX: Children's Hospital Oxford
- CVS: Chorionic Villus Sampling FBC: Full Blood Count
- DGH: District General Hospital
- EDD: Estimated Date of Delivery
- OHTC: Oxford Haemophilia and Thrombosis Centre
- IBDs: Inherited Bleeding Disorders
- ID: Iron deficiency
- IDA: Iron Deficiency Anaemia
- ISTH BAT score: International Society for Thrombosis and Haemostasis Bleeding Assessment Tool score
- LMP: Last Menstrual Period
- TXA: Tranexamic Acid

There should be clear communication between the maternal medicine centre (Silver Star at the John Radcliffe Hospital) as well as the patient's local obstetrician, local shared care haematologist, Adult haemophilia centre and Paediatric haemophilia team.

We strongly encourage that all babies who are confirmed to be affected by a bleeding disorder or who potentially have a bleeding disorder are delivered in Oxford under the Silverstar Unit where the Adult and Paediatric Haemophilia Team and Specialist Haemostasis lab are available 24/7.

This guideline recognises diversity in diagnosis and management of inherited bleeding disorders. Whilst the common hereditary bleeding disorders (haemophilia A and B, and Von Willebrand disease) do not show a predilection for a particular ethnicity, international data suggest that outcomes for these patients do vary with ethnicity and socioeconomic deprivation. Ethnic variation in von Willebrand factor levels can also affect diagnosis. Achieving equality of health outcomes requires identification of barriers, and biases. It needs targeted action to overcome specific inequalities, discrimination, and marginalisation experienced by certain groups and individuals as highlighted in the Equality Act 2010. Many sources highlight the consequences of complex pregnancy and potential morbidity and mortality. Women and birthing people from Black and minority ethnic backgrounds (BME), along with those with severe and multiple disadvantages are more likely to die in pregnancy compared to white women or those who do not have a disadvantage.
We aim to provide equal and accessible care to all service users with a focus on underrepresented groups or those with multiple disadvantages. This can be achieved with the use of collaborative, specialist, multi-agency input and support. This includes early intervention and effective communication between services. The vision is to ensure exceptional quality care for all through equitable access, experience, and optimal outcomes.