# The Yorkshire and The Humber Maternal Medicine Network

# Haematology Principles

## Purpose and Scope

The Yorkshire and Humber (Y&H) region have come together to develop the Y&H Maternal Medicine Network (MMN). The aim of the MMN is to align care across the region and to reduce inequalities in care to women/ birthing people with complex medical conditions. The purpose of this document is to outline the key principles that should be adopted when caring for women/ birthing people with Haematological conditions. This document should be used in conjunction with the 'Conditions for Consideration of Referral to the Maternal Medicine Centre' document, which can be found at www.maternalmedicine.org.uk This document has been approved by governance at both LTHT and STH. This document can either be used as a standalone document following ratification by adopting Trusts or can be used as a reference to incorporate into Trust guidance.

Findings from national reports, such as MBRRACE, have shown that those who are from a Black, Asian or Mixed Ethnic Background or those who have a severe mental illness or live in the most deprived neighbourhoods, are at higher risk of poorer physical health and care outcomes in addition to their medical condition/s. Therefore, it is important to consider the person's individual needs, circumstances and wider determinants of health and offer reasonable adjustments to address these to ensure improved equity of access, support, and care for individuals. The perinatal period adds further complexity, therefore please ensure you consider mental health needs of the patient and refer to your local perinatal mental health service appropriately. Additional information can be found at www.everymummatters.com

## Haematology Maternal Medicine Network Referral Pathway:

| Care led by Maternal Medicine Centre | Review, advice & guidance from MMC | Local expertise |
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| Haemophilia A/B:<br>▪ Fetus affected by moderate to severe haemophilia (or not known whether fetus affected)<br>▪ Carrier of haemophilia with low levels Factor VIII/IX | Haemophilia A/B:<br>▪ Haemophilia carrier (refer carriers of haemophilia as early as possible)<br>▪ Partner of pregnant patient with Haemophilia A/B | |
| Von Willebrands:<br>▪ Type 2 & 3 VWD<br>▪ Type 1 VWD if VWF not normalised | Von Willebrands:<br>▪ Type 1 VWD: VWF normalised in pregnancy | |
| Any bleeding disorder already under care in MMC, or likely to require haemostatic support antenatally or peripartum to reduce haemorrhage risk (including severe platelet disorders) | Mild bleeding disorder, or partner of patient with mild bleeding disorder (platelet function defect, other mild coagulation factor deficiency such as Factor XI deficiency) | |
| | Current ITP and platelets <75 | ▪ Gestational thrombocytopenia<br>▪ Historical ITP and platelets >75 |
| ▪ Antithrombin deficiency<br>▪ Thrombotic Antiphospholipid Syndrome | Inherited thrombophilia with previous VTE | Inherited thrombophilia (no previous VTE, not antithrombin deficiency) |
| | Current extensive VTE or new VTE > 36/40 gestation | ▪ Current or previous VTE event<br>▪ Obstetric antiphospholipid syndrome |
| Sickle cell disease | | Sickle cell trait |
| Transfusion-dependent thalassaemia | ▪ Non-transfusion dependent thalassaemia<br>▪ Thalassaemia trait and Hb <75 | Alpha/beta thalassaemia trait |
| Active haematological malignancy | Myeloproliferative disorders | Previous treated haematological malignancy |
| TTP requiring treatment | TTP in remission | |
| PNH | Rarer red cell disorders already under MMC care | Thrombocytosis |

# Venous thromboembolism (VTE) prophylaxis and treatment

| VTE thromboprophylaxis principles of care |
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| All women should have a documented risk assessment for VTE at booking, on admission to hospital, in labour or immediately postnatal, after birth and if any additional problems present during pregnancy. |
| Assessment and management of VTE risk should be as per the RCOG Greentop Guideline: 'Reducing the Risk of Venous Thromboembolism During Pregnancy and the Puerperium' |
| The risks of VTE should be discussed with all women and individual recommendations discussed. |
| If a woman develops bleeding whilst on low molecular weight heparin (LMWH) the treatment should be stopped, and expert haematological advice sought. |

| VTE thromboprophylaxis - pre pregnancy care |
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| Women with previous VTE should be offered pre pregnancy counselling to develop a prospective plan for pregnancy. |

| VTE thromboprophylaxis- Antenatal care |
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| Women should be offered thromboprophylaxis as per the RCOG Greentop Guideline: 'Reducing the Risk of Venous Thromboembolism During Pregnancy and the Puerperium' |
| Antenatal thromboprophylaxis for those with previous VTE should begin as early in pregnancy as practical. |
| Trusts should ensure that there is a process in place to enable early access to first trimester thromboprophylaxis for those requiring it (MBRRACE 2024) |
| Women with previous VTE associated with antithrombin deficiency or antiphospholipid disease (who will often be on long term anti-coagulation) should be referred to the MMN and urgent advice sought from a haematologist regarding LMWH dosage. |
| Women with a previous VTE and an inherited thrombophilia should be referred to the MMN. |
| All inpatient pregnant women should be prescribed LMWH unless there is a specific contraindication. |

| VTE thromboprophylaxis - Intrapartum care |
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| Women receiving antenatal LMWH should be advised that if they have any vaginal bleeding, or once labour begins, they should not inject any further LMWH. They should be reassessed on admission to hospital and further doses prescribed by medical staff. |

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| Regional techniques should be avoided until at least 12 hours after the previous prophylactic dose of LMWH. |
| Women receiving antenatal LMWH having an elective caesarean section should receive a thromboprophylactic dose of LMWH on the day prior to delivery. On the day of delivery any morning dose should be omitted and the operation performed that morning. |
| Women receiving thromboprophylaxis should be prioritised within the induction of labour pathway to reduce time off anticoagulation (MBRRACE 2021). |

| **VTE thromboprophylaxis - Postnatal care** |
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| Women should be offered thromboprophylaxis as per the RCOG Greentop Guideline: 'Reducing the Risk of Venous Thromboembolism During Pregnancy and the Puerperium' |
| The first thromboprophylactic dose of LMWH should be given as soon as possible after delivery, provided there is no post-partum haemorrhage and regional anaesthesia has not been used. |
| LMWH should not be given for 4 hours after spinal anaesthesia/epidural catheter removal and an epidural catheter should not be removed within 12 hours of the most recent LMWH injection. |
| In women who have additional persistent risk factors, such as prolonged admission, wound infection or surgery in the puerperium, thromboprophylaxis should be extended for up to 6 weeks or until the additional risk factors are no longer present. |

| **Acute Venous Thromboembolism – Principles of Care** |
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| Any woman with symptoms and/or signs suggestive of VTE should have objective testing performed as soon as possible and treatment with LMWH until the diagnosis is excluded by objective testing. |
| All units should have an agreed protocol for the diagnosis of suspected VTE during pregnancy based on the RCOG Greentop Guideline: 'Thromboembolic Disease in Pregnancy and the Puerperium:Acute Management'. |
| Women with proven VTE in pregnancy should receive input from a haematologist. |
| Women with extensive VTE or VTE > 36 weeks gestation should be referred to the MMC. |
| Women on therapeutic LMWH should have individualised plan for labour and delivery. |
| All units need to have clear pathways/treatment protocols for women who are unstable with pulmonary embolism including joint care between physicians, obstetricians, critical care and radiology. |

## Sickle Cell Disease

| Sickle cell - Pre pregnancy care |
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| Advise preconception counselling with a sickle cell specialist haematologist and an obstetrician |
| Recommend folic acid 5mg od for 3/12 prior to conception |
| Screen for end-organ damage: echocardiogram, BP and proteinuria, liver and renal function, retinal screening, screening for iron overload |
| Review medications and switch to pregnancy-safe options if required. |
| Ensure woman is taking penicillin prophylaxis and that immunisations are up to date. |
| Check partner status – if a carrier provide information about pre-implantation genetic diagnosis and prenatal testing. |

| Sickle cell - Antenatal care |
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| Offer partner testing if not known ideally by 10 weeks to allow for 1st trimester diagnosis +/- termination. |
| Refer to the Maternal Medicine Centre. |
| Review screening for end organ damage, vaccinations and medications. Check FBC, U&E, LFT, urinary PCR, ferritin and extended RBC phenotype. Monitor FBC monthly from 20 weeks gestation. |
| Actively manage nausea and vomiting to reduce risk of dehydration and sickle cell crises. |
| Recommend aspirin 150mg od from 12 weeks gestation. |
| Recommend folic acid 5mg od, penicillin prophylaxis and Vitamin D supplementation throughout pregnancy. |
| Start LMWH prophylaxis at 28 weeks or earlier if additional risk factors are present. |
| Check urine culture monthly. |
| Arrange anaesthetic assessment. |
| Offer monthly growth scans from 24 weeks gestation. |
| Plan timing and mode of delivery by 36 weeks gestation. |
| Recommend delivery at 38-40 weeks gestation. |

| Sickle cell- Intrapartum care |
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| Advise hospital birth at a unit able to manage complications of sickle cell disease. |
| Sickle cell disease should not influence mode of delivery. |
| Management in labour should be overseen by a multi-disciplinary team including midwives, senior obstetrician, anaesthetist and haematologist. |
| Recommend continuous fetal monitoring. |
| Monitor oxygen saturations, keep warm and well hydrated, offer effective analgesia (avoid pethidine) and have a low threshold to start antibiotics if any suspicion of infection. |
| Cross-match blood if atypical antibodies present. Need for blood transfusion should be determined by haematologist and experienced obstetrician - blood should be matched for extended phenotype and should be CMV negative. |

| Sickle cell - Postnatal care |
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| Offer early testing for sickle cell disease if baby is at high risk. |
| Ensure adequate hydration and maintain SATs ≥ 94%. |
| Recommend LMWH for 6 weeks postnatally. |
| Provide contraception advice. |

| Sickle cell – Management of acute crises |
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| Provide analgesia (avoid pethidine, can use NSAIDs between 12 and 31 weeks) |
| Screen for infection and acute chest syndrome (ACS). Prescribe antibiotics if indicated. |
| Assess need for fluids and oxygen |
| Give LMWH prophylaxis. |
| Arrange early MDT review by both haematology and obstetrics. |
| After 28 weeks gestation recommend daily CTG assessment and consider ultrasound for fetal growth assessment if not performed in the preceding 2 weeks. |

## Thrombocytopenia:

| Thrombocytopenia Antenatal Care |
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| In women with platelets 101-149 monitor once in each trimester if they remain> 100 (booking, 28 weeks and 36 weeks) |
| Women with falling platelets or known low platelets (<100) need an antenatal clinic review to establish cause (ITP/inherited/Gestational). Consider requesting local haematology input. |
| Refer women with platelets <75 to the MMC for advice alongside local haematology input. |
| Arrange anaesthetic referral for women with platelets <80. |
| Consider neonatal alert if confirmed ITP or unclear diagnosis with platelets <100. |
| LMWH is contraindicated in women with platelets <75 unless under guidance from haematology. |
| Women with thrombocytopenia do not require additional fetal surveillance. |
| Thrombocytopenia is not an indication for early delivery. |
| All women with platelets <100 should have a delivery plan which includes suitability for fetal scalp electrode (FSE), instrumental delivery, regional analgesia/anaesthesia and a postpartum management plan. |

| Thrombocytopenia intrapartum care |
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| Recommend hospital birth for all women with platelets <100. Recommend individualised birth setting for women with platelets 101-149. |
| When admitted in labour perform FBC and G&S |
| Women in active labour with platelets < 100 require IV access |
| Ensure anaesthetic team aware when woman with platelets <100 is admitted in active labour. |
| Women with thrombocytopenia do not require continuous monitoring unless additional risk factors or individualised care plan. |
| Advise senior obstetrician presence at delivery if a woman with platelets <100 requires instrumental delivery or full dilatation caesarean section. |
| In women with known or suspected ITP or platelets <80 check fetal platelet count via cord blood. |

| Thrombocytopenia Postnatal care |
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| Ensure appropriate haematology follow up arranged if required |
| VTE thromboprophylaxis can be given (if indicated) provided platelets are >75. |
| Intramuscular vitamin K should be avoided until the neonatal platelet count is known. |

## Bleeding Disorders

| Bleeding disorders - Pre pregnancy care |
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| In women with a bleeding disorder, female carriers of haemophilia or women with a partner affected by Haemophilia, advise preconception counselling with a Bleeding Disorders Haematologist and an obstetrician (refer to the Maternal Medicine Centre). |
| Counsel regarding inheritance, preimplantation and antenatal testing options (NIPT, CVS, early and late amniocentesis). |
| Establish non-pregnant baseline VWF or factor levels if the woman has von Willebrands disease or is a carrier of Haemophilia. |

| Bleeding disorders - Antenatal care |
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| Refer to the Maternal Medicine Centre. Antenatal care should be provided by a multidisciplinary team including haematologists, obstetricians and midwives. |
| Women with bleeding disorders (including WVD and female carriers of Haemophilia) are at increased risk of bleeding with invasive procedures, termination, miscarriage and at the time of delivery. Check relevant factor level at booking, before any invasive procedure, in the third trimester and if any bleeding symptoms. A plan for testing and haemostatic support during any invasive procedures should be made in advance. Factor levels of > 0.5 iu/ml are usually required for invasive procedures. |
| Carriers of severe Haemophilia A or B should be offered NIPT for fetal sex from 9 weeks gestation at the local unit. Confirm fetal sex at 20/40 anatomy USS. |
| Carriers of severe Haemophilia A or B with a male baby by NIPT should be offered CVS at 11-14 weeks gestation |
| Carriers of severe haemophilia with a male baby who decline early prenatal screening should be offered amniocentesis between 34-36 weeks to inform mode of delivery. |
| External cephalic version should be avoided in fetuses potentially affected by Type 2 or 3 VWD, male fetuses affected by, or potentially affected by, Haemophilia A or B and in female fetuses who are obligate or possible carriers of severe Haemophilia B. |
| For women with bleeding disorders a delivery plan should be made in the antenatal period detailing place and mode of delivery, any delivery restrictions, haemostatic support for the women, anaesthetic considerations, management in the postnatal period (including VTE prophylaxis if indicated) and testing for the baby. |
| Complete paediatric alert. |

| Bleeding disorders - Intrapartum care |
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| Advise hospital birth. The MMC will make recommendations regarding place of birth in conjunction with the woman's wishes. |
| If delivery restrictions are recommended they usually include avoidance of fetal scalp electrode, fetal blood sampling, mid-cavity forceps and ventouse. Specific variations may be advised in the delivery plan. |

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| In women with VWD and carriers of Haemophilia factor levels of > 0.5 iu/ml are required for delivery, neuroaxial anaesthesia and intramuscular injections. (Specific advice may given in the case of Type 2 and 3 VWD). |
| Tranexamic acid, DDAVP and Factor IX may be recommended for intrapartum haemostasis: timing and dosage should be detailed in the delivery plan along with recommendations for fluid management if DDAVP is recommended. |
| For other factor deficiencies and bleeding disorders an individualised delivery plan should be made including advice about haemostatic support and neuroaxial anaesthesia. |
| Offer active management of the third stage. |

| **Bleeding disorders - Postnatal care** |
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| In women with VWD, or carriers of Haemophilia A and B, specific guidance about factor testing and haemostatic treatment in the postnatal period should be included in the delivery plan, as well as guidance about VTE prophylaxis if indicated. |
| A plan for diagnostic testing of the neonate following delivery, including cord blood sampling, should be included in the delivery plan. Plans regarding further investigations (e.g. intracranial ultrasound), factor treatment and follow up will be made in conjunction with the neonatal team.<br>Women with bleeding disorders should be encouraged to report any excessive bleeding in the postnatal period. |

# References

RCOG Greentop Guideline: ‘Reducing the Risk of Venous Thromboembolism During Pregnancy and the Puerperium’

RCOG Greentop Guideline: Bleeding Disorders in Pregnancy

RCOG Greentop Guideline: ‘Thromboembolic Disease in Pregnancy and the Puerperium:Acute Management’.

British Society Haematology: Guidelines for the Management of Sickle Cell Disease in Pregnancy. Aug 2021

MBRRACE 2024, 2021, 2015

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