



**GUIDELINES FOR
MANAGEMENT OF PREGNANCY
AND DELIVERY IN WOMEN
WHO ARE EITHER CARRIERS
OR PATIENTS WITH BLEEDING
DISORDERS**

Australian Haemophilia Centre Directors'
Organisation

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Disclaimer

This document is a general guide to appropriate practice, to be followed subject to the clinician's expert judgment and the patient's preference in each individual case. While the guidelines are derived from the current evidence based literature, they are designed to provide a practical guide and thus are purposely more succinct, but lack extensive references to the literature. Such a literature review has been recently provided by the National Blood Authority (Evidence-based clinical practice guidelines for the use of recombinant and plasma-derived Factor VIII and Factor IX products – <http://www.nba.gov.au>). The guidelines are based on a consensus opinion at the time of compilation (June 2007). These guidelines will be reviewed in 2010.

Users of these guidelines are encouraged to contact the Australian Haemophilia Centre Directors' Organisation if they have comments to make. These comments will inform the revision process.

Users can contact the Australian Haemophilia Centre Directors' Organisation by writing to:

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1. Physiological response expected in pregnancy in women with bleeding disorders

Factor VIII and vWF antigen usually increase significantly during pregnancy in women with type 1 vWD and in haemophilia A carriers. They reach their maximum between 29 and 35 weeks. Factor IX and XI levels usually do not change significantly during pregnancy. Because factor levels are not predictable during pregnancy, repeat testing during the third trimester is recommended. After delivery, factor levels usually return to baseline after days to weeks, but may drop earlier and should be monitored carefully if baseline levels would provide inadequate haemostasis. It should be noted women with normal factor levels may still be carriers.

2. Management of pregnancy and delivery

2.1. During pregnancy

Usually no specific therapy is required during pregnancy.

If the mother is a haemophilia carrier, or there is a history of haemophilia in the family, genetic counseling is desirable.

In preparation for delivery, factor levels should be measured during the third trimester (usually at 32-34 weeks)

Invasive procedures can be performed if the factor level is normal, otherwise replacement therapy may be required.

The patient should be managed jointly with a 'high risk' Obstetric Unit and HTC (haemophilia treatment centre).

2.2. Labour and delivery

An inherited bleeding disorder is not an indication per se for delivery by Caesarean section and vaginal delivery is preferred.

The ideal hospital for delivery is where there is both a 'high risk' Obstetric Unit and HTC.

Treatment in bleeding disorders will need to be individualized and specific treatment protocols should be followed (see references).

A normal factor level in the mother (or quantitative/functional von Willebrand's studies in vWD) is desired for delivery.

Factor replacement should be given to the mother as close to delivery as possible. (N.B. This does not normalize the baby's factor level)

Vacuum extraction is contraindicated. Forceps should be avoided where possible, as should foetal scalp blood sampling and scalp electrodes.

DDAVP should not be used prior to delivery due to risk of fluid retention, hypotension, uterine contractions and premature labour.

2.3. Postpartum Care

It is generally recommended to keep factor levels in the normal range for 3 to 4 days after a vaginal delivery and up to 7 days after a Caesarean section.

2.4. Obstetric Anaesthesia

Epidural anaesthesia should only be considered in close consultation with the anaesthetist. Congenital bleeding disorders are associated with an increased risk of spinal haematoma. Other forms of analgesia may be preferred. Epidural anaesthesia is generally not recommended for use in severe type 2 or 3 VWD.

There are no guidelines adequately covering epidural/spinal anaesthesia in patients with bleeding disorders.

Coagulation studies should be maintained in the normal range for the duration of catheter placement and for 12 hours (mild bleeding disorder) to 24 hours (moderate to severe bleeding disorder) after catheter removal.

2.5. Postpartum haemorrhage

Active management of third stage should be practised.

Early postpartum haemorrhage, associated with low factor levels at the time, should be managed by factor replacement therapy or DDAVP in VWD Type 1 or Haemophilia A. DDAVP is not effective in Haemophilia B or FXI deficiency and some types of vWD. DDAVP is not contra-indicated during lactation.

Should late postpartum haemorrhage occur, tranexamic acid, oral contraceptives and longer-term, the Levonorgestrel-releasing intrauterine device are first-line therapy for its management. Tranexamic acid is safe in breast feeding mothers and is category B1 in pregnancy.

2.6. Management of neonate

If the baby is at risk of having a severe bleeding disorder, blood samples should be taken for factor levels.

Cord blood testing is controversial particularly in haemophilia B (Factor IX clotting activity in cord blood in a normal-term newborn is lower than in adults (mean: ~30%); thus, the diagnosis of haemophilia B can be established in an infant with activity less than 1%, but is equivocal in an infant with moderately low activity).

In general it may provide a rapid and accessible opportunity to exclude severe disease but results should be confirmed with peripheral blood testig. In addition, testing should always be repeated at six months of age.

In general, intramuscular injections should be avoided.

Vitamin K is often given orally or subcutaneously to avoid the risk of intramuscular haematoma. If intramuscular injection is performed this should only ever be used in experienced HTC's. Pressure should be applied for 10 minutes to the injection site.

A trans-fontanel ultrasound should be performed soon after birth in babies 'known to be affected' with a severe bleeding disorder.

Even in neonates 'known to be affected' with a severe bleeding disorder, 'prophylactic' factor replacement should not be given due to any potential risk of inhibitor development. The use of 'prophylactic' rFVIIa has also not been shown to improve clinical outcomes.

All infants, including those with bleeding disorders, should be immunised for hepatitis B. Prophylactic factor replacement therapy in patients with severe haemophilia is not usually required or encouraged due to the potential of immune system stimulation and increased risk of inhibitors.

Neonates with a known inherited bleeding disorder should be registered at a HTC.

2.7. Management of bleeding in neonates

Neonates known (or suspected) to have haemophilia and who have evidence of either intracranial bleeding or severe bleeding elsewhere, should receive immediate factor replacement with recombinant factor VIII or IX.

If the type of haemophilia is unknown then both factor VIII and IX should be given until confirmation with specific factor levels is obtained.

Neonates with severe haemophilia A or B and severe bleeding require 100% plasma factor levels which can be achieved by giving doses of recombinant

factor VIII of 75IU/kg and factor IX 150IU/kg respectively. Factor levels should be maintained in the normal range for at least 7-10 days and therapy co-ordinated by an HTC and haemophilia specialist. In patients with early product exposure consideration of continuing regular factor replacement prophylaxis should be given as it may reduce the risk of inhibitors.

Guidelines for the care of a child with haemophilia are not covered in this document.

3. References

Bolton-Maggs P. Factor XI deficiency and its management. www.wfh.org.

Canadian Hemophilia Society: Women with inherited bleeding disorders.
www.hemophilia.ca

Demers, C., Derzko, C., David, M. & Douglas, J. (2005). Gynaecological and obstetric management of women with inherited bleeding disorders. *Journal of Gynaecology Canada, July*, 707 – 718.

Haljamae H. (1996). Thromboprophylaxis, coagulation disorders, and regional anaesthesia. *Acta Anaesthesiol Scand, 40 (8 Pt 2)*, 1024-40.

Horlocker TT, Wedel DJ, Benzon H, et al. (2003). Regional anesthesia in the anticoagulated patient: defining the risks (The second ASRA consensus conference on neuraxial anesthesia and anticoagulation). *Reg Anest Pain Med, 28*,172-197.

Kadir, R. & Lee CA. (2005). Obstetrics and gynecology: hemophilia. In C.A Lee, E.E Berntorp & W.K Hoots, (eds). *Textbook of Hemophilia*. Blackwell Publishing, pp. 249-56.

Lee CA, Chi C, Pavord SR, et al. (2006). The obstetric and gynaecological management of women with inherited bleeding disorders – review with guidelines produced by a taskforce of UK Haemophilia Doctors' Organisation. *Haemophilia; 12*, 301-336.

National Blood Authority: Evidence-based clinical practice guidelines for the use of recombinant and plasma-derived Factor VIII and Factor IX products.
www.nba.gov.au