Detailed Management of the VWD Patient in Pregnancy

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- A careful personal and family bleeding history is important.
- In patients with suspected vWD test at 34-36 weeks (or earlier if preterm delivery is likely).
- Request von Willebrand's screen (record blood group).
- *Remember: normal levels do not exclude a diagnosis of vWD.*
- Avoid epidural anaesthesia (see below).

Treatment Options

These include either DDAVP or plasma derived factor VIII (CSL AHF). Do not use recombinant factor VIII, plasma, or cryoprecipitate.

Management at Delivery

- If the Factor VIIIc parameters are normal at 34-36 weeks manage expectantly but with a high index of suspicion for postpartum haemorrhage.
- If the Factor VIII levels are unusual or fail to normalise (e.g =50%) consider prophylaxis:-

Either

- DDAVP (0.3 mcg/kg) given following clamping of umbilical cord.

CAUTION: DDAVP is an antidiuretic agent and can cause hyponatraemia. Care with I.V. fluid replacement.

Or

- Plasma derived factor VIII (CSL AHF 250 units/reconstituted bottle). This should be used if there is a hstory of significant bleeding with a previous delivery.
- Approximately 3-4 bottles administered by slow I.V. push.

Post Partum Haemorrhage

In the event of postpartum bleeding, where prophylaxis has not been given, treatment will be:-

- DDAVP (0.3 mcg/kg) by I.V. infusion (diluted in crystalloid over 20 minutes).
- *CAUTION: DDAVP is an antidiuretic agent and can cause hyponatraemia. Care with I.V. fluid replacement.*

I bleeding is significant plasma derived factor VIII (CSL AHF – 3-4 bottles by slow I.V. push) should be given.

Spinal-Epidural Anaesthesia for Labour and Delivery

The majority of patients with type I von Willebrand's disease show normalisation of factor VIII parameters in pregnancy. If factor VIII and von Willebrand factor levels are normal at 36 weeks and there is no reason to manage the delivery in any special way but rather to follow normal routine practice, including use of episiotomy.

The issue of whether or not spinal or epidural anaesthesia is used depends on the willingness of the anaesthetist. It should be acknowledged that if coagulation parameters have normalised with a normal platelet count, there is minimal, if any, bleeding risk associated with an atraumatic lumbar puncture. The potential issue relates to medico-legal risk.

If the anaesthetist is conversant with the risks, and following full discussion with the patient, it may be acceptable to proceed with epidural or spinal anaesthesia if required (particularly for lower segment Caesarean section) if:

- A coagulation screen (including an assessment of platelet function (PFA)) and platelets are normal at the time that procedure is planned.
- That the lumbar puncture is performed by an experienced anaesthetist with an atraumatic technique.
- Consider the use of DDAVP at the time of the needle/catheter insertion (particularly if a traumatic tap occurs) and possibly at the time of removal of the catheter. Best discussed with an haematologist.
- Strict neurological evaluation following the procedure with early intervention if there are any signs of a haematoma.

The Infant

Von Willebrand's disease is an autosomomal dominantly inherited condition with variable penetrance (approximate one third of at risk infants will inherit the condition).

- Avoid invasive foetal monitoring (eg. scalp vein sampling) when possible. Care with instrumental deliveries.
- Give vitamin K at birth.

Infants are not routinely tested unless they have unexplained bleeding problems.

Miscarriage

- Bleeding during pregnancy requires urgent obstetric consultation.
- Patient with an early miscarriage may require no additional treatment. If there is a need for intervention to remove retained products or prolonged bleeding treatment with DDAVP should be considered.
- Patients with a personal history of miscarriage or bleeding during pregnancy may require more frequent monitoring of von Willebrand factor parameters during pregnancy.