BC Women's Hospital Department of Anesthesia

Peripartum management of a parturient with type 1C (clearance) von Willebrand disease

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Introduction

Typical type 1 von Willebrand Disease (vWD) self corrects in the third trimester in response to placental estrogen, and responds adequately to DDAVP

Type 1C (clearance) vWD is a subtype not previously described in obstetric anesthesia literature, characterised by:
1.Increased clearance of vWF
2.Failure to self-correct in pregnancy
3.Short-lived response to DDAVP

Case History

36 year old G3 P0 patient referred to clinic at 32 weeks due to history of type1 vWD. Previous DDAVP response testing, and factor levels through pregnancy are shown in the tables

Poor response to DDAVP and inadequate third trimester self-correction of vWF and FVIII were identified, prompting referral to the adult bleeding disorders clinic

A care-plan was formulated in advance for peripartum hemostatic management, including administration of VWF/FVIII concentrate

Alternatives to neuraxial techniques (including remifentanil PCA and general anesthesia) were discussed in the event that vWF/FVIII replacement was not possible (e.g. in the emergency setting)

Summary of peripartum management

75 IU/kg vWF/FVIII concentrate was administered intravenously prior to epidural placement (requested at 4cm dilation)

After 4h, a second 75 IU/kg dose was administered.
The epidural catheter was removed after an uneventful vaginal delivery, following discussion with hematology

Postpartum, 35 IU/kg vWF/FVIII concentrate was administered at 12h, 24h, 36h, followed by three doses of 45 IU/kg daily

1g IV tranexamic acid was given at delivery, then 1.5 g PO daily for 10 days

Type 1C von Willebrand Disease (vWD) accounts for ~5% of all type 1 vWD

In this subtype, von Willebrand factor (vWF) and Factor VIII (FVIII) fail to correct in the 3rd trimester, and response to DDAVP is short-lived

vWF/FVIII concentrate is likely to be required prior to peripartum neuraxial placement

Laboratory results

Table 1: Factor levels through pregnancy

Lab Parameter	Normal Range	Baseline (Pre-pregnancy)	19 weeks	32 weeks	35 weeks
vWF antigen(U)	> 0.5	0.1	0.15	0.21	0.31
FVIII activity (U)	0.5-1.5	0.26	0.27	0.34	0.35
vWF:RCo (U)	> 0.5	0.14	N/A	N/A	0.23
vWFpp:vWF antigen ★	0.9-1.62	13.0	N/A	N/A	N/A

★ A raised vWF propeptide (vWFpp): vWF antigen ratio is a marker of accelerated vWF clearance (this test may be requested by hematology to support diagnosis of type 1C vWD

Table 2: Pre-pregnancy response to DDAVP testing (0.3 mcg/kg IM)

Time after DDAVP	FVIII Activity (U/ml)	vWF antigen (U/ml)	vWF:RCo (U/ml)
Reference range	0.5 – 1.5	> 0.50	> 0.50
Baseline	0.26	0.14	0.14
1 hour	1.10	0.70	0.59
4 hours	0.44	0.23	0.28
24 hours	0.20	0.11	0.13

KEY POINTS

- Type 1 vWD usually self-corrects in pregnancy, but vWF and FVIII levels should always be checked in the 3rd trimester
- vWF/FVIII concentrate may be required if vWF and FVIII remain low and previous response to DDAVP is inadequate
- Prescription of vWF/FVIII concentrate is specific to individual patients and should be guided by a hematologist, as genotypes and phenotypes variable considerably
- Multidisciplinary care including a specialist bleeding disorders team is always advisable in atypical vWD, such as type 1C