



PAEDIATRIC ACUTE CARE GUIDELINE

Von Willebrand Disease

Scope (Staff):	All Emergency Department Clinicians
Scope (Area):	Emergency Department

This document should be read in conjunction with this DISCLAIMER
<http://kidshealthwa.com/about/disclaimer/>

Von Willebrand Disease

Also see guideline [Management of a Child with a Bleeding Disorder](#)

Background

- Von Willebrand Disease is a common autosomal dominant disorder, with three subtypes.
- Von Willebrand Disease usually presents with easy bruising, mucosal bleeding, post-operative bleeding or post traumatic bleeding. Menorrhagia and post-partum haemorrhage are common presentations in females.

Treatment for von Willebrand Disease is usually with:

- DDAVP (Desmopressin) 0.3 microgram/kg diluted in 0.9% saline and given IV over 20 minutes for the common type-1 patients.
- Patients with type-2 disorders (functional deficiency) and the rare type-3 disorder (homozygous disorder) will require factor replacement therapy with plasma-derived factor VIII (Biostate).
 - Recombinant factor VIII **does not** contain von Willebrand factor
 - Supplies of plasma-derived factor VIII (Biostate) are kept in the fridge in Transfusion Medicine (Haematology laboratory)
- **All presentations to the Emergency Department should be discussed with the on-call Haematologist before any treatment is instituted**
- Please refer to the [Haematology Transfusion Medicine Protocols](#) for further information

Indications for admitting a patient with an underlying bleeding disorder:

- Suspected intracranial haemorrhage
- Persistent mouth bleeding not responding to factor replacement therapy




and antifibrinolytic therapy

- Persistent haematuria
- Severe persistent epistaxis
- Undiagnosed abdominal pain
- Suspected psoas haemorrhage
- Bleeding into hip or inguinal area
- Compartmental syndrome such as forearm bleeding
- Bleeding into neck
- Tonsillar haemorrhage
- Tight soft tissue bleeds

References

External Review: Catherine Cole (Paediatric and Adolescent Oncologist/Haematologist) - July 2015

This document can be made available in alternative formats on request for a person with a disability.

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