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Pathology Newsletter
April 2023

Von Willebrand Disease and your patient

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Introduction

Von Willebrand Disease (VWD) is a coagulation disorder which is caused by either low levels or dysfunctional Von Willebrand Factor (VWF).

The VWF is an early coagulation protein which interacts with platelets in concert with factor VIII in the initial clotting process. Deficiency or abnormal VWF therefore manifests as with prolonged and / or excessive bleeding.

Although men and women are equally affected, women present more frequently due to complications of menstruation and / or childbirth.

Three large subgroups have been described.

Type 1 is caused by low levels of VWF whereas type 3 by absent VWF. Type 2 is subdivided as 2A (VWF is the wrong size), 2B (the timing of VWF attachment to platelets are wrong), 2M (no platelet binding occurs) and 2N (no f.VIII binding occurs).

Clinical manifestations that should prompt suspicion of VWF

1. Frequent or hard-to-stop nose bleeds.
a. Spontaneous bleeding.
b. Occurs five or more times per year.
c. Lasts for longer than 10 minutes.
d. Requires packing or cautery to stop.

2. Easy bruising
a. Frequent bruising with little or no trauma.
b. Bruise is larger than a R5 coin.
c. Forms a raised lump.

3. Heavy menstrual bleeding.
a. Requiring to change pad and / or tampon every 1 - 2 hours.
b. Menstruations lasting more than 7 days.
c. Flooding or gushing blood.
d. Passing of clots the size of grapes or larger.
e. Subsequent diagnosis of anaemia.

4. Prolonged bleeding following childbirth, surgery, injury or dental work.

Diagnosis

Initial screening

The initial testing should include a full blood count with a coagulation profile and fibrinogen level (table 1).

Table 1. Initial testing and findings used in the diagnosis of VWD

Test	Physiological measurement	Findings
Full Blood Count	Determine whether there are other causes of coagulation disorders like thrombocytopaenia.	Typically normal features of iron deficiency anaemia may be present.
APTT	Function of coagulation factors VIII, IX, XI and XII.	Prolonged due to functional f.VIII deficiency but may be normal in mild cases.
PT	Function of coagulation factors II, V, VII and X.	Normal.
Fibrinogen	Co-factor in coagulation, where a deficiency may lead to coagulation disorders.	Normal.

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Follow up testing

It is important to note that testing should best be performed in the absence of excess physiological stress, pregnancy or infection as these functional tests are highly affected.

In order to confirm a diagnosis of VWD and its type, the following tests need to be performed and interpreted by a pathologist and / or haematologist:

1. Factor VIII clotting activity.
2. VWF antigen.
3. Ristocetin cofactor.
4. Von Willebrand factor multimer analysis.
5. Platelet aggregation tests.

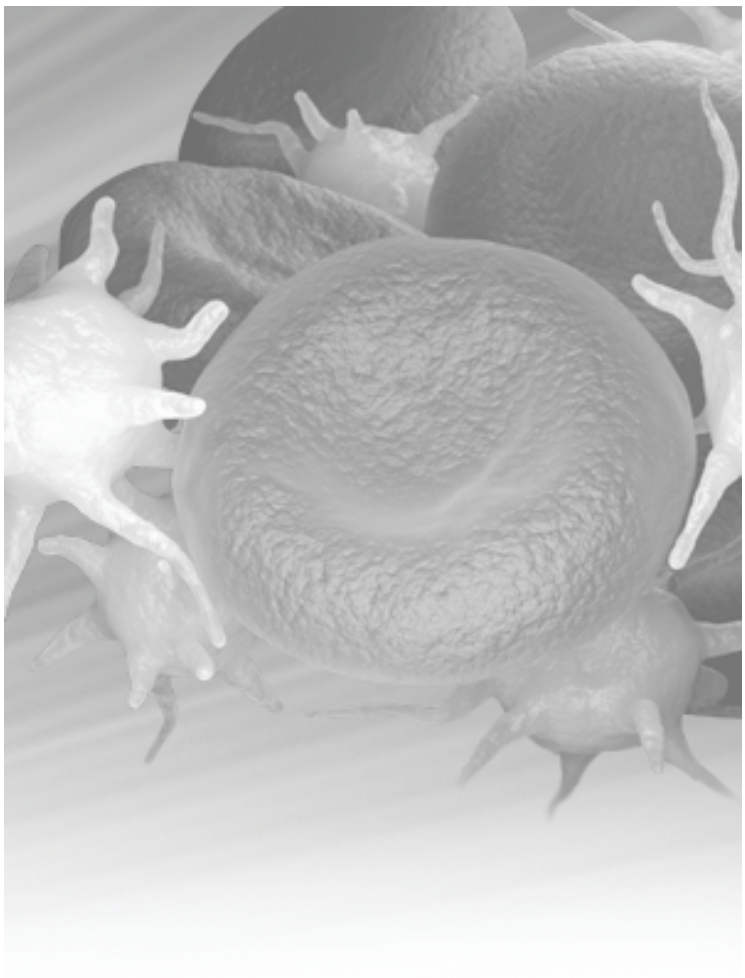
Depending on the different results for these assays, the diagnosis and typing can be performed.

Treatment options

1. Desmopressin acetate injections.
2. Desmopressin acetate nasal spray.
3. Factor replacement containing both VWF and f.VIII.
4. Antifibrinolytics.
5. Oral contraception.

References

1. For more patient information, access the CDC website at https://www.cdc.gov/ncbddd/vwd/-facts.html



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