von Willebrand disease

Von Willebrand disease (vWD) is an inherited autosomal disorder or, rarely, an acquired bleeding disorder.

The disease is caused by the quantitative deficiency or dysfunction of von Willebrand factor (vWF).

Von Willebrand factor has two main functions in haemostasis. It is an adhesion protein that is essential for platelet-plug formation, diverting circulating platelets to the sites of vascular injury, particularly through larger multimers; and it forms a noncovalent complex with factor VIII in plasma, thereby protecting it from being inactivated and cleared.

Patients with von Willebrand disease may notice frequent or prolonged epistaxis, easy bruising, menorrhagia, and excessive or prolonged bleeding after surgery.

von Willebrand disease

Type 1	Partial quantitative deficiency of qualitatively normal vWF
Type 2	Qualitative defects of vWF
2A	Defective platelet-dependent vWF functions, associated with lack of larger multimers
2B	Heightened platelet-dependent vWF functions, associated with lack of larger multimers
2M	Defective platelet-dependent vWF functions, not associated with lack of larger multimers
2N	Defective vWF binding to factor VIII
Type 3	Severe or complete deficiency of vWF

Laboratory tests

Coagulation studies (a normal result does not exclude vWD)

Platelet Function Analysis (PFA)

Blood group and von Willebrand studies, which include:

- Factor VIII
- vWF Antigen
- vWF Ristocetin cofactor activity
- vWF Collagen Binding activity

(Medicare Australia requires each of these tests to be ordered individually)

and not commonly:

- vWF Multimer analysis
- Factor VIII binding assay.

Treatment

Desmopressin (DDAVP) with initial trial to assess response.
Factor VIII concentrates.