

Table 1 Summary of the main tests used in the investigation of von Willebrand disease

Test	Abbreviation	What the test measures
A. Routine blood tests		
Complete or full blood counts/morphology	CBC or FBC	White cell, red cell, and platelet counts and other indices. In the context of VWD, platelet count and morphology are potentially useful. For example, a low platelet count may be seen in type 2B or platelet type VWD, or may instead reflect immune or idiopathic thrombocytopenia or a platelet defect
Prothrombin time	PT	Sensitive to deficiencies in factors II, V, VII, X. Will usually be normal in VWD unless additional comorbidity is present (e.g., vitamin K deficiency)
Activated partial thromboplastin time	aPTT	Sensitive to deficiencies in factors II, V, VIII, IX, X, XI, XII. Will sometimes be abnormal in VWD (if FVIII is low), but will be normal in most cases of VWD
Thrombin time/fibrinogen	TT/Fib	Should be normal in VWD, unless additional comorbidity is present
B. Typical VWD screening test panel		
Factor VIII coagulant activity	FVIII:C	The level of functional FVIII. Usually by one stage clotting assay based on a modified aPTT; sometimes by chromogenic assay (several manufacturers/suppliers)
VWF antigen	VWF:Ag	The level of VWF (both functional and not). Historically by ELISA, now mostly by latex immunoassay (several manufacturers/suppliers); sometimes by chemiluminescence (one manufacturer/supplier).
VWF glycoprotein Ib-binding activity	VWF:GPIbB	Various methods (see below)
VWF ristocetin cofactor	VWF:RCo	A VWF:GPIbB performed using platelets and ristocetin to measure platelet agglutination (several manufacturers/suppliers). Historically, the original VWF activity assay.
VWF GPIb binding using recombinant GPIb	VWF:GPIbR	A VWF:GPIbB performed using latex or magnetic particles, recombinant GPIb, plus ristocetin to respectively measure latex agglutination or chemiluminescence based events (one manufacturer/supplier). A modern alternative to VWF:RCo.
VWF GPIb binding using recombinant mutated GPIb	VWF:GPIbM	A VWF:GPIbB performed using latex (commercial method; one manufacturer/supplier) or ELISA (not yet commercialized), recombinant mutated gain of function GPIb (but no ristocetin) to respectively measure latex agglutination or ELISA color generation. Another modern alternative to VWF:RCo.
C. Additional tests for VWD investigation		
VWF–collagen-binding activity	VWF:CB	Primarily performed by ELISA (a large number of manufacturers/suppliers), and increasingly by chemiluminescence (one manufacturer/supplier). We use the latter, and also include this test in our screening panel.
VWF factor VIII-binding activity	VWF:FVIII B	Primarily performed by ELISA (one manufacturer/supplier; or using in house/laboratory developed methods).
Ristocetin induced platelet aggregation/ agglutination	RIPA	Performed by platelet agglutination/aggregation (one manufacturer of ristocetin, but distributed by several suppliers)
VWF multimers	VWF:mult	Performed by agarose gel electrophoresis (one commercial semiautomated method; otherwise in house/laboratory developed methods)

Abbreviations: CBC, complete blood count; ELISA, enzyme-linked immunosorbent assay; GPIb, glycoprotein Ib; FBC, full blood count; FVIII, factor VIII; RIPA, ristocetin-induced platelet agglutination/aggregation; VWD, von Willebrand disease; VWF, von Willebrand factor.