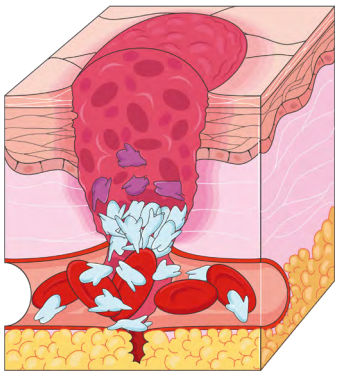
NORDIC BIOMARKER

von Willebrand Factor

von Willebrand Factor and Its Functions

von Willebrand factor (vWF) is a large protein, ranging in size from dimers to large multimers, where the largest multimers have the most activity. The size of vWF is regulated by the protein ADAMTS13.

During vascular damage, subendothelial collagen is exposed, which vWF binds to. During this binding, vWF undergoes a conformational change which allows binding sites to become exposed. These binding sites allows the activation, adherence and aggregation of platelets, which is essential in the primary haemostasis.



vWF plays an important part in the primary haemostasis, enabling platelets and red blood cells to form plugs in wounds.
Image credit: Servier Medical Art

vWF acts as a carrier molecule to Factor VIII, preventing its degradation. In doing so, vWF acts as a procoagulant in the secondary haemostasis.

von Willebrand Disease

von Willebrand disease (vWD) is the most common inherited bleeding disorder, affecting up to 1% of the population. The disease is caused by decreased level or function of von Willebrand factor (vWF). The signs and symptoms of vWD include frequent nosebleeds or nosebleeds that are hard to stop, easy bruising, bleeding in the gums and heavy menstrual bleeding. In severe cases (such as in Type 3 vWD), joint and muscle bleeding can be seen.

von Willebrand disease is divided into three types.

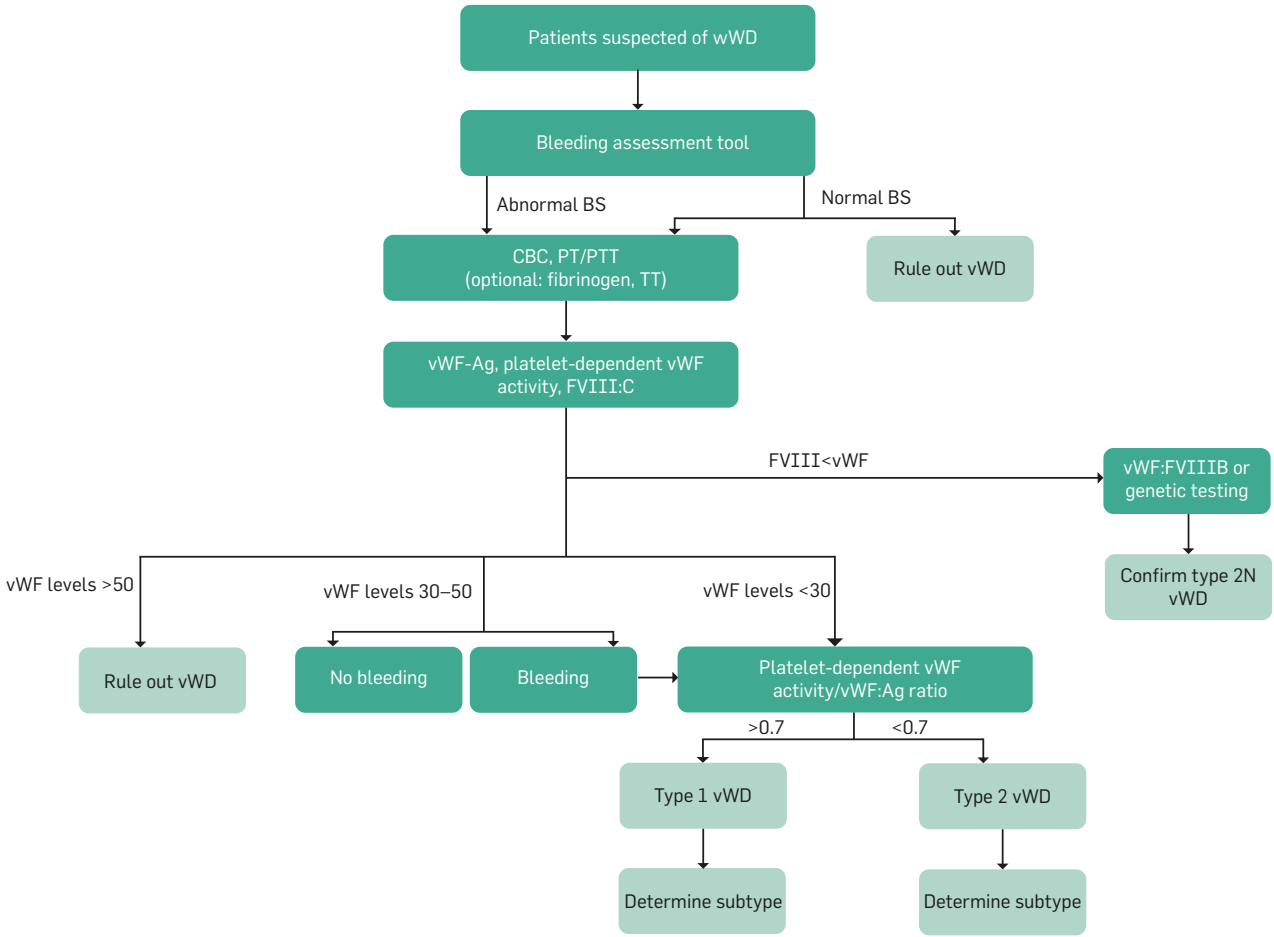
Type 1	**Type 2**	**Type 3**
70–80% of cases	Around 20% of cases	Less than 5% of cases
Quantitative deficiency Lower levels of vWF than normal.	Qualitative deficiency Normal levels of vWF, but the protein is dysfunctional. Four subtypes.	Absence of vWF

Treatment

There are several treatments for vWD. Oral contraceptives, desmopressin, tranexamic acid or a von Willebrand factor concentrate might be administered depending on the severity of the disease.

Testing for Von Willebrand Disease

When von Willebrand disease is suspected in a patient, the level and activity of vWF must be determined to set a diagnose. The diagnosis process is complicated and also takes clinical symptoms and family history into account. The level of vWF (called vWF antigen) in circulation, the activity of vWF and the activity of factor VIII is assayed. The ratios between the results of these assays will confirm or rule out von Willebrand's disease and aid in the diagnosis of the specific type.



Simplified and reduced algorithm for the diagnosis of vWD. Source of recommendation: ASH ISTH NHF WFH 2021 guidelines on the diagnosis of von Willebrand disease

A Closer Look: MRX Green vWF Antigen

MRX Green vWF Antigen consists of vWF antigen specific monoclonal antibodies coupled to sub-micron sized polystyrene particles.

When the reagent is exposed to a plasma sample containing vWF antigen, the particles will agglutinate, giving rise to increased light-scattering. When exposed to the appropriate wavelength of light (500–700 nm), the increase in measured turbidity, or light-scattering, is proportional to the amount of vWF Antigen in the sample. The assay is calibrated against the WHO international standard and correlates well to existing assays on the market.