

Von Willebrand Disease

Von Willebrand disease happens when you don't have enough or the right kind of a special protein that helps stop bleeding. This protein acts like glue to hold platelets in place at the site of an injury. Without enough good glue, bleeding takes longer to stop.

THE ONE-LINE TRUTH

Von Willebrand disease is a failure of the glue that helps platelets stick to injured blood vessels, making bleeding last longer than it should.

The chain — how it unfolds

1. Von Willebrand factor is absent, deficient, or structurally abnormal — why: Inherited gene mutations prevent normal production, storage, or release of functional VWF protein from endothelial cells and platelets.
2. Platelets cannot adhere to injured blood vessel walls — why: Without VWF to bridge platelets to exposed collagen, platelets slide past the injury site instead of sticking and forming the initial hemostatic (bleeding-stopping) plug.
3. Primary hemostasis fails and bleeding time prolongs — why: The first phase of clotting depends on a platelet plug forming within 1-3 minutes; without VWF this takes 10-30 minutes or longer, allowing continued oozing from cuts and injuries.
4. Factor VIII levels drop due to lack of carrier protein — why: VWF normally protects factor VIII from rapid enzymatic breakdown; without this protection, factor VIII is cleared from blood in 2 hours instead of 12, dropping levels to 5-40% of normal.
5. Secondary hemostasis is also impaired — why: Low factor VIII weakens the intrinsic coagulation pathway (one of the clotting cascades), reducing thrombin generation and fibrin mesh formation needed to strengthen the clot.
6. Mucosal and soft tissue bleeding becomes frequent — why: Areas with high blood flow and friable (easily damaged) tissue—nose, mouth, GI tract, uterus—bleed easily and persistently because weak clots break apart under normal blood flow pressure.

7. Chronic or recurrent bleeding leads to iron deficiency anemia — why: Repeated bleeding episodes, especially heavy menstrual periods (menorrhagia) in women, cause ongoing iron loss exceeding dietary intake, dropping hemoglobin (oxygen-carrying protein) to 7-10 g/dL or lower.
8. Rare but life-threatening bleeding (hemorrhage) occurs with major trauma or surgery — why: In Type 3 disease or severe Type 2 variants, complete absence of functional VWF means no primary hemostasis at all, leading to uncontrolled bleeding into joints (hemarthrosis), muscles, or brain after injuries that would be minor in healthy individuals.

Spot it — what you see, and why

- Easy bruising: Low von Willebrand factor (VWF, a clotting protein) prevents platelets (clotting cells) from sticking to blood vessel walls, so even minor bumps cause bleeding under the skin
- Frequent nosebleeds lasting more than 10 minutes: Nasal blood vessels are fragile and without adequate VWF to plug tiny breaks, bleeding continues longer than normal
- Heavy menstrual bleeding requiring pad or tampon change every 1-2 hours: Uterine (womb) blood vessels cannot form stable clots during menstruation without sufficient VWF, so the endometrium (lining) sheds with excessive blood loss
- Prolonged bleeding after dental extraction or surgery: Surgical wounds need platelet plugs held together by VWF; deficiency allows the plug to fall apart and bleeding to restart hours or days later
- Blood in stool or black tarry stools: Gastrointestinal (stomach and intestine) bleeding from angiodysplasia (abnormal blood vessels) or ulcers cannot clot properly without VWF
- Joint swelling and pain: Hemarthrosis (bleeding into joints) occurs in severe type 3 VWD where factor VIII (another clotting protein carried by VWF) is also very low

Red flags — act now

- Hemarthrosis (joint bleeding) or deep muscle a pooled collection of blood (hematoma) suggesting severe type 3 VWD with very low factor VIII
- Postpartum bleeding (hemorrhage) requiring transfusion or ICU admission
- Signs of hypovolemic shock (tachycardia, low blood pressure (hypotension), altered mental status) from acute blood loss
- New onset bleeding in older adult suggesting acquired von Willebrand syndrome from lymphoma, myeloproliferative disorder, or aortic narrowing (stenosis)

Treat it — in this order

1. Desmopressin (DDAVP) given intravenously, subcutaneously, or intranasally before procedures → Stimulates release of stored VWF from endothelial cells (blood vessel lining) into circulation, raising VWF and factor VIII levels 3-5 fold within 30-60 minutes
2. VWF/factor VIII concentrate infusion for major bleeding or surgery → Directly replaces missing VWF and factor VIII proteins, restoring normal clotting within minutes; used when DDAVP is ineffective or contraindicated
3. Tranexamic acid oral or intravenous → Blocks plasmin (enzyme that breaks down clots), stabilizing clots already formed in mucous membranes like mouth, nose, and uterus for 6-8 hours per dose
4. Hormonal contraceptives (combined oral pills, levonorgestrel IUD, or depot medroxyprogesterone) for heavy menstrual bleeding → Thins the endometrium (uterine lining) and reduces menstrual blood loss by 40-50 percent; estrogen in combined pills also raises VWF levels by 20-30 percent
5. Avoidance of antiplatelet drugs (aspirin, NSAIDs, clopidogrel) and anticoagulants unless absolutely necessary → Prevents additional impairment of platelet function and clotting cascade, reducing bleeding risk in daily life

Your top priorities at the bedside

1. Assess for active bleeding and bleeding risk before any procedures or surgeries — why: Von Willebrand disease (VWD) is a bleeding disorder caused by low or poor-working von Willebrand factor (a protein that helps blood clot). Patients bleed longer than normal, so we must check bleeding history and current bleeding sites before invasive procedures to prevent life-threatening bleeding (hemorrhage).
2. Monitor menstrual bleeding patterns in female patients and adolescents — why: Heavy menstrual bleeding (menorrhagia) is often the first sign of VWD in women and girls. Excessive periods can cause anemia (low red blood cells) and fatigue. Early recognition helps start treatment and prevent complications like iron deficiency.
3. Teach patients to avoid aspirin and NSAIDs like ibuprofen — why: Aspirin and nonsteroidal anti-inflammatory drugs (NSAIDs) stop platelets (clotting cells) from working properly. In VWD patients who already have clotting problems, these medicines increase bleeding risk significantly. Acetaminophen is safer for pain.