The Blood Project

UNDERSTANDING VON WILLEBRAND DISEASE TYPE 2

A brief guide for patients with a functional form of von Willebrand disease

Von Willebrand disease type 2 is a form of von Willebrand disease in which the body makes von Willebrand factor, but the protein does not work normally. Von Willebrand factor plays a key role in blood clotting by helping platelets stick to injured blood vessels and supporting normal clot formation. Type 2 von Willebrand disease is less common than type 1 and varies in severity. Many people live full, active lives, but symptoms and bleeding risk can be more variable and may not match routine blood test results.

What is von Willebrand disease type 2?

Type 2 von Willebrand disease occurs when von Willebrand factor is present, but does not function properly.

In this form, the issue is not how much protein is present, but how well it works. As a result, bleeding can last longer than expected, particularly during surgery, dental work, or injury, even when von Willebrand factor levels appear near normal.

Type 2 is best understood as a **functional problem**, rather than a problem of low quantity.

Why does it happen?

Type 2 von Willebrand disease is usually inherited and results from a genetic change that affects the structure or behavior of von Willebrand factor.

Because the protein is made but does not work as it should, standard blood tests that measure the *amount* of von Willebrand factor may look reassuring, while tests that measure *function* show abnormalities. This mismatch is a common source of confusion.

As with other forms of von Willebrand disease, test results can change with stress, illness, inflammation, hormones, pregnancy, and aging. These changes are expected and do not mean the diagnosis is uncertain.

Are there different forms of type 2?

Yes.

Type 2 von Willebrand disease includes several subtypes, which differ in how the protein's function is altered. These subtypes help doctors understand bleeding risk and choose the safest treatment approach.

Patients do not need to memorize subtype names. What matters most is that your care team understands your specific pattern and plans accordingly.

Does it cause symptoms?

Sometimes.

Symptoms vary widely and may include:

- easy bruising
- frequent or prolonged nosebleeds
- heavy menstrual bleeding

- bleeding that takes longer than expected to stop
- prolonged bleeding after dental work or surgery

In type 2, symptoms may feel **out of proportion to blood test results**, because the protein is present but not functioning well. Symptoms deserve attention even when numbers appear near normal.

Is type 2 von Willebrand disease dangerous?

For most people, it is manageable and does not shorten life expectancy.
However, bleeding risk in type 2 can be **less predictable** than in type 1, particularly around procedures, childbirth, or major injury. Serious bleeding is uncommon, but planning is especially important.
Knowing about the diagnosis ahead of time allows doctors to prevent bleeding rather than respond after it occurs.

How your doctor evaluates it

Doctors diagnose type 2 von Willebrand disease by looking at **patterns and function**, not a single number. Evaluation often includes:

- blood tests that measure how well von Willebrand factor works
- tests that compare function to amount
- related clotting tests
- a careful bleeding history
- repeat testing to confirm consistent patterns

Because von Willebrand factor behaves dynamically, one normal or borderline test does not rule out type 2.

What Is the treatment?

Many people with type 2 do not need daily treatment.
Treatment is used when bleeding risk is higher, such as before surgery, dental work, or childbirth.
Options may include:

- medicines that help stabilize clots
- von Willebrand factor replacement during high-risk situations
- treatment plans tailored to subtype and bleeding history

Some treatments used in type 1 are **not effective or not appropriate** in certain forms of type 2 (for example, medications that work well in type 1 may not be used in some type 2 subtypes). This is why accurate diagnosis and advance planning matter.

When should I contact my doctor?

You should contact your doctor if bleeding patterns change or if you are planning a procedure. Reach out if you experience:

- new or worsening bleeding
- heavy menstrual bleeding that interferes with daily life or causes anemia
- bleeding that is harder to stop than expected
- upcoming surgery, dental work, or pregnancy
- questions about medications that affect bleeding

Seek urgent medical care for severe bleeding, head injury, severe abdominal pain, chest pain, or severe shortness of breath.

What is the usual plan going forward?

People with type 2 von Willebrand disease are often followed by a hematologist, sometimes in partnership with a primary care clinician. Follow-up focuses on:

- understanding your specific bleeding pattern
- planning ahead for procedures and life events
- reassessing symptoms over time

The goal is **anticipation and preparation**, not restriction. With appropriate planning, most people manage type 2 safely and effectively.

Key points to remember

- **functional problem:** von Willebrand factor is present but does not work normally
- **numbers can be misleading:** function matters as much as amount
- **symptoms may not match labs:** bleeding history is essential
- **planning is critical:** especially for procedures and childbirth
- **most people do well:** with awareness and individualized care