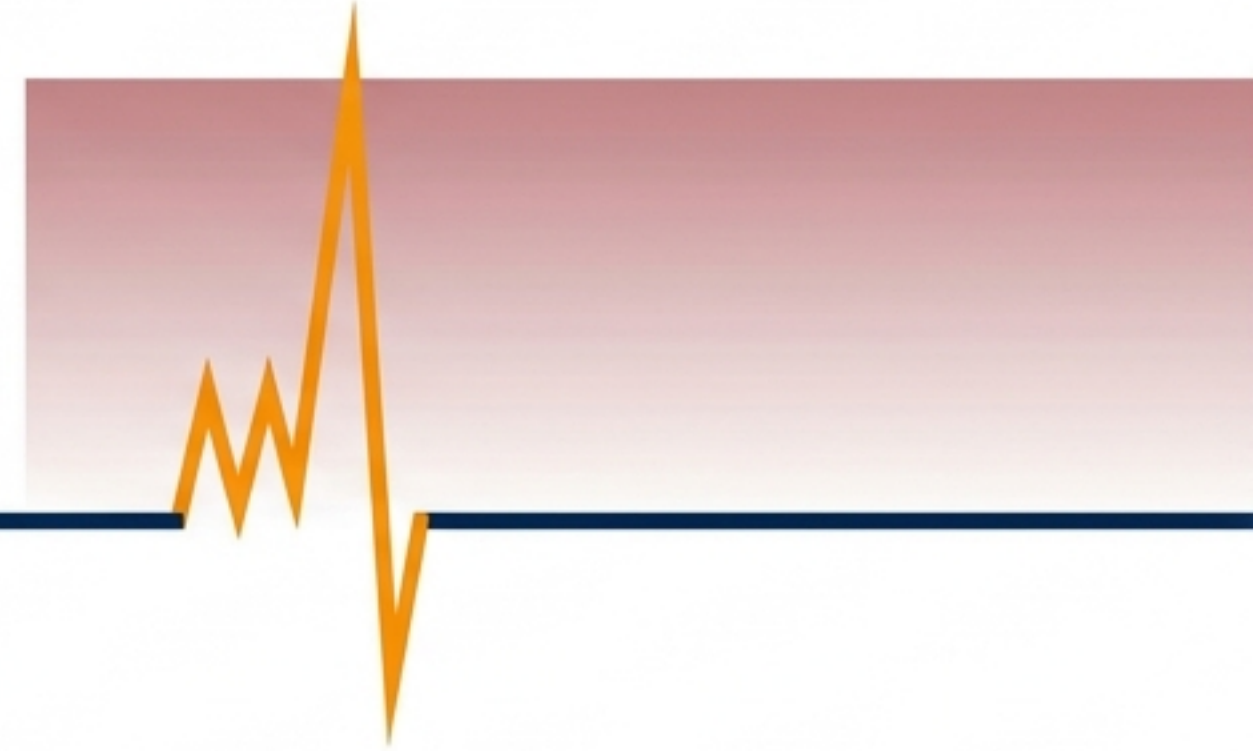
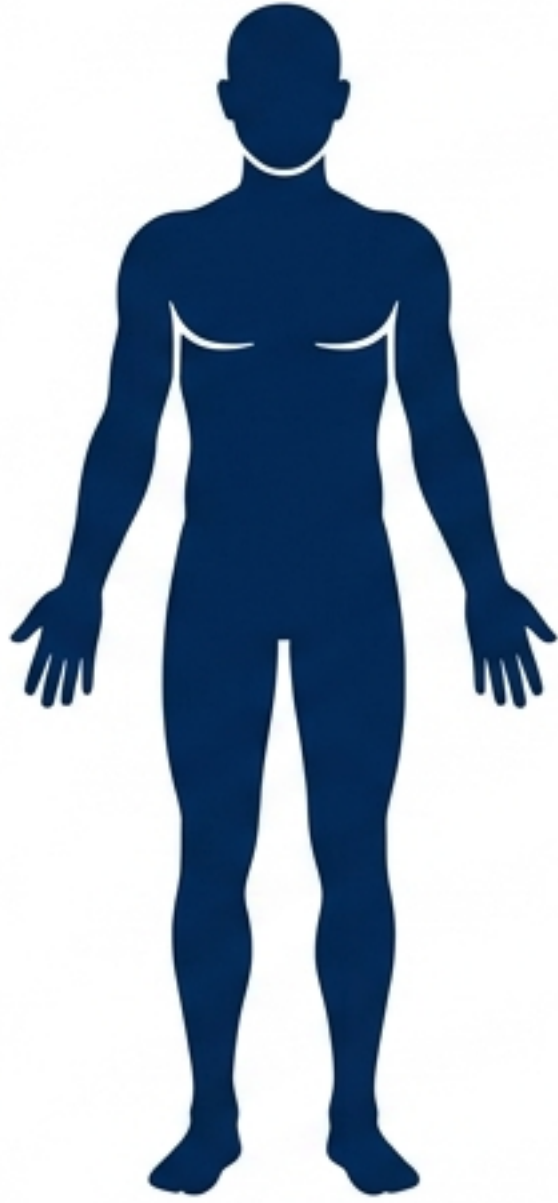


Biologie Biology Stress



The Clinical Presentation of
von Willebrand Disease

By William Aird



Many patients with VWD do not bleed every day.
They look well between hemostatic challenges.

The question is not simply:
Does this patient bleed?

**The better question is:
When has this
patient's hemostatic
system been tested,
and what happened
when it was?**

VWD bleeding is not random. It is VWF biology made visible under stress.

VWF: THE DUAL FUNCTION MICRO-DIAGRAM

THE TETHER (PRIMARY HEMOSTASIS)



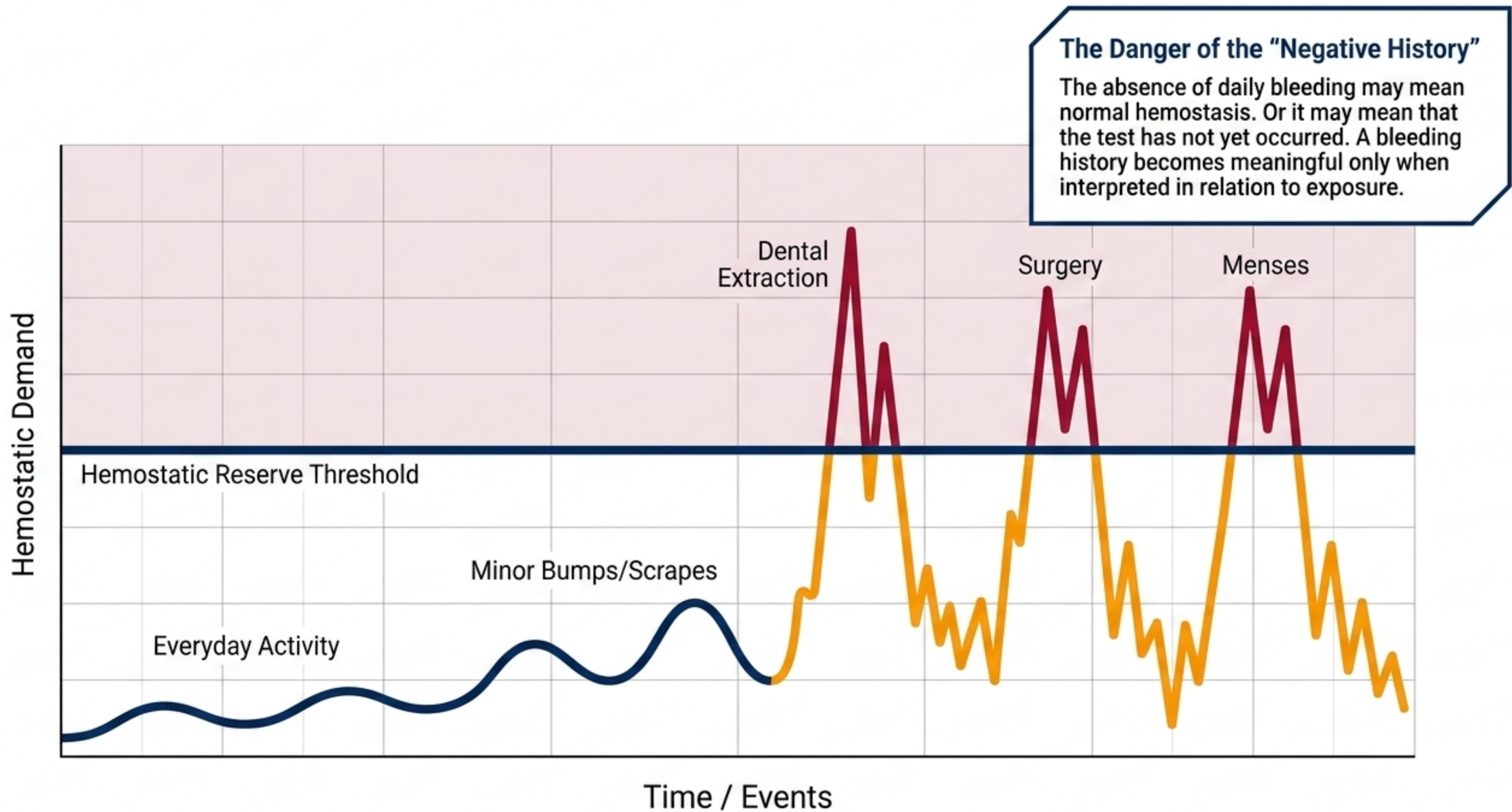
Supports Platelet Adhesion: VWF **tethers platelets** to sites of vascular injury, acting as the critical initial plug—especially under high shear stress.

THE CARRIER (COAGULATION CASCADE)

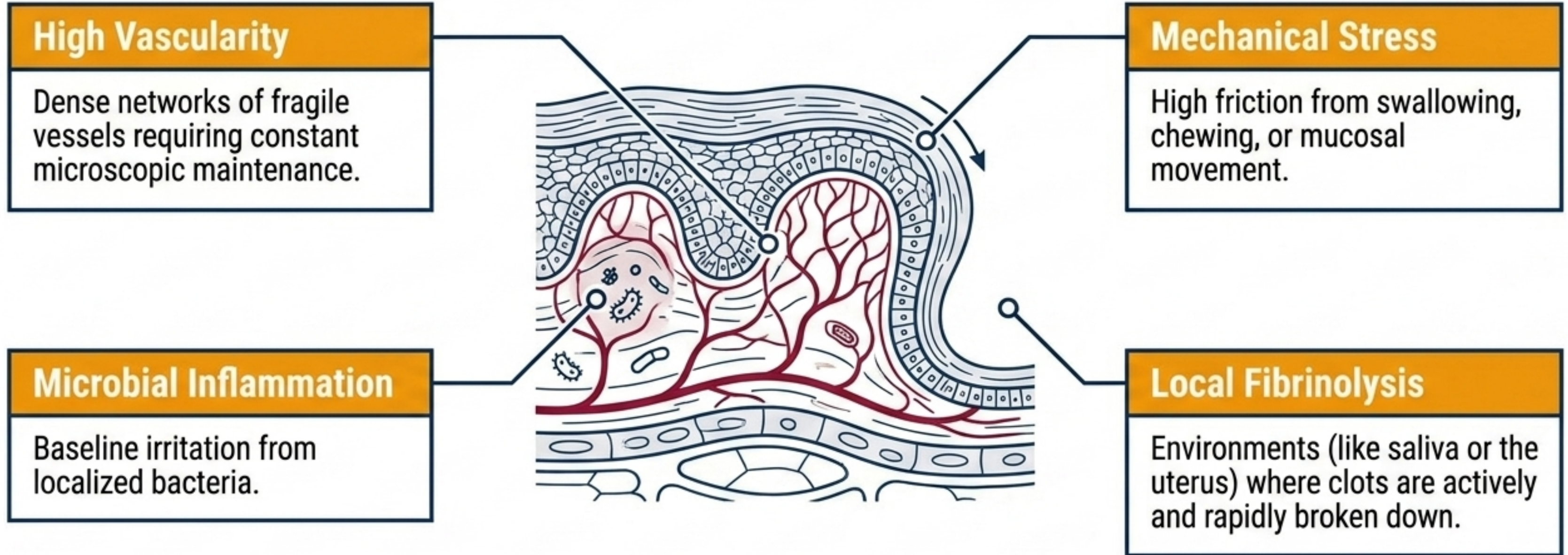


Stabilizes Factor VIII: VWF acts as a **protective carrier protein**. Without it, FVIII is cleared too quickly, disrupting the secondary coagulation cascade.

When local hemostatic demand exceeds baseline reserve in either of these functions, bleeding occurs.



Why Mucosal Surfaces Reveal VWD



In VWD, the problem is not that bleeding starts in strange places. It is that **bleeding in common, hemostatically demanding places** behaves abnormally.

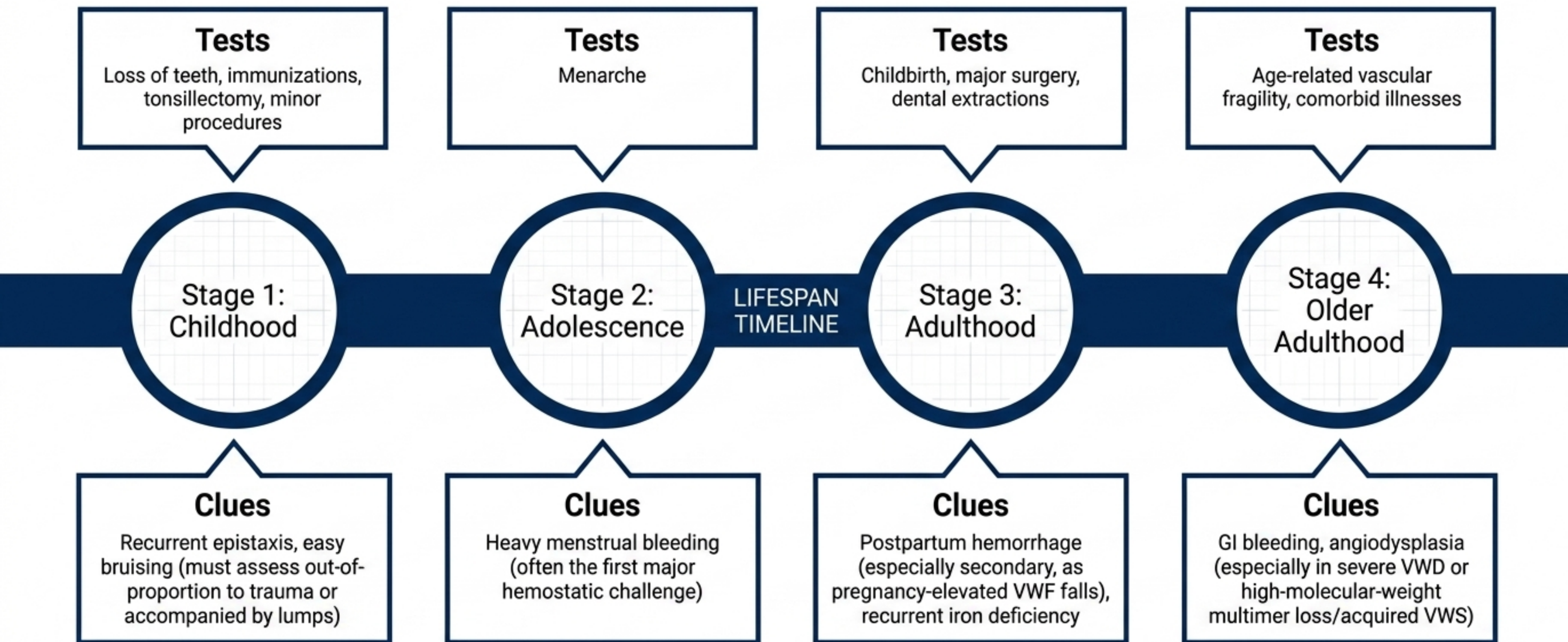
The “True Negative” vs. “False Reassurance” Matrix

The Tested Patient (True Negative)	
Profile	Adult who has undergone surgery, dental extraction, childbirth, or severe trauma
Outcome	No abnormal bleeding
Diagnostic Weight	 Highly reassuring. The hemostatic reserve has held under intense load.

The Untested Patient (False Reassurance)	
Profile	Child, pre-menarchal adolescent, man without prior procedures, or adult who avoided surgery
Outcome	No abnormal bleeding
Diagnostic Weight	 Not reassuring. The system’s limits remain unknown. Low scores on Bleeding Assessment Tools (BATs) are uniquely misleading here.

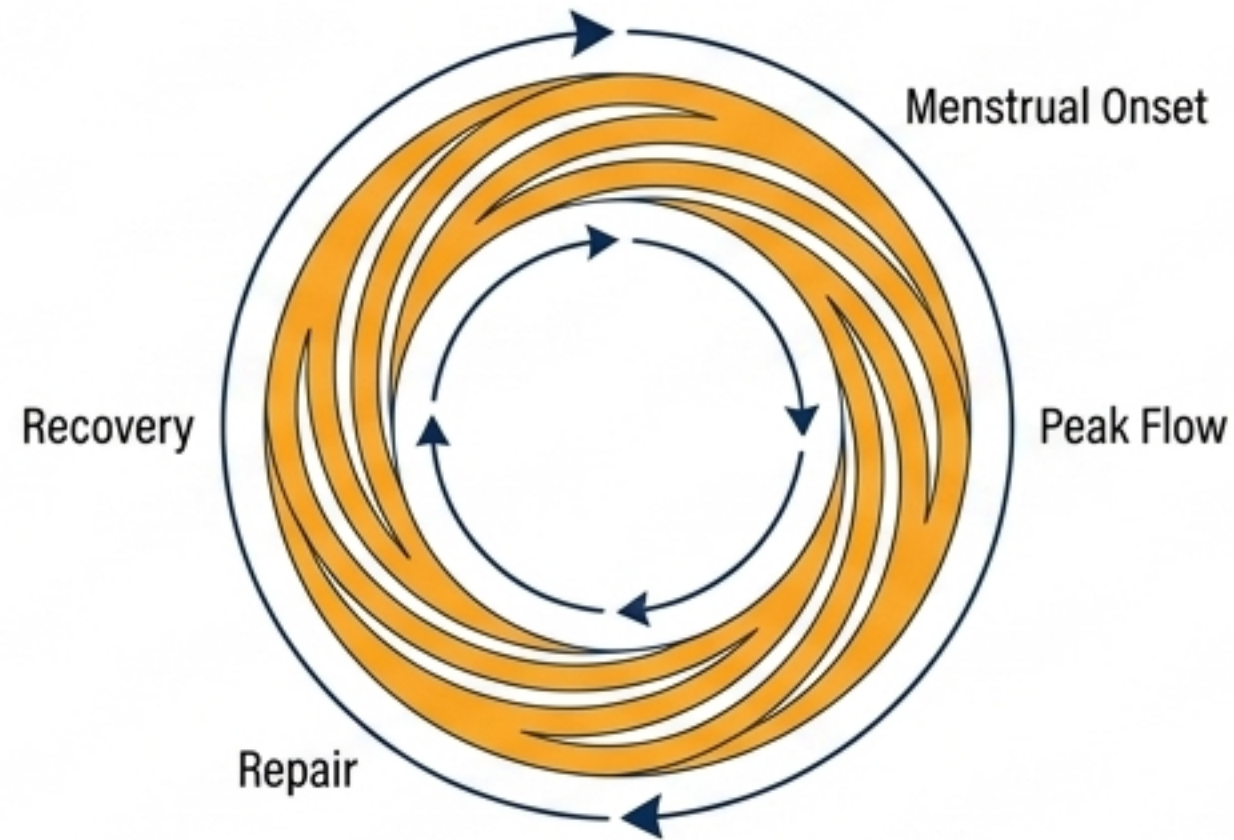
A surgical history is not just a list of operations. It is a record of how the patient handled hemostatic stress.

The VWD Clinical Journey: A Lifespan Timeline



The Ultimate Stress Test: Heavy Menstrual Bleeding

Cyclical Hemostatic Challenge



In menstruating patients, the uterus acts as the **first repeated, high-demand test of primary hemostasis**. Clues to system failure include:

- | | | |
|--|---|-----------------------------------|
| ▪ Passage of clots larger than 1 inch. | ▪ Changing pads/tampons more than hourly. | ▪ Nighttime changes and flooding. |
|--|---|-----------------------------------|

Ferritin: The Bleeding Indicator



Ferritin is part of the bleeding story.

A normal hemoglobin does not exclude clinically important iron loss. Iron deficiency is the measurable consequence of chronic mucosal bleeding.

The Intraoperative Timeframe (The Water Line)

The Massive Hidden Risk:
Delayed, recurrent, or persistent
postoperative oozing.

Dental Extraction

Combines vascularity, saliva,
local fibrinolysis, and
mechanical stress. Initial clot
may form, then break down.

The Tip: Immediate surgical hemorrhage
(rarely the primary presentation).

Tonsillectomy

High-risk mucosal wound.
Airway involvement means
delayed bleeding during the
postoperative healing phase
is uniquely dangerous.

**Delayed mucosal bleeding should never be dismissed simply because
initial intraoperative hemostasis appeared satisfactory.**

When VWD Looks Like Hemophilia

Phenotype Matrix		
	Impaired Tether (Standard Phenotype)	Impaired Carrier (Hemophilia-like Phenotype)
Mechanism	Reduced VWF quantity/function for platelet adhesion.	VWF is nearly absent or fails to bind and stabilize Factor VIII.
Subtypes	Mild Type 1, many qualitative subtypes.	Severe Type 3, Type 2N VWD.
Clinical Pattern	Mucocutaneous bleeding (epistaxis, bruising, heavy menses).	Deep tissue bleeding, muscle hematomas, hemarthroses.

Insight: A disproportionately low Factor VIII level does not automatically equal Hemophilia A. Sometimes the problem is not FVIII production—it is VWF failing to carry it.

VWD Diagnosis: The Limitations of Routine Screening



Routine Screening Tests
Do Not Exclude VWD

Platelet Count: Typically normal.

PT: Normal.

aPTT: Often normal because FVIII
may only be mildly reduced.

PFA-100/200: Abnormal in severe
disease, but unreliable for ruling out
mild/moderate VWD.



Require Focused VWF
Panels in Context

VWF Antigen

Platelet-dependent VWF Activity

Factor VIII Activity

When the clinical bleeding phenotype is convincing, the next step is not reassurance based on normal screening tests. It is focused testing.



Bleeding severity rarely matches a single VWF level perfectly because the level is only one quarter of the equation.

THE PATIENT FILE

PATIENT:	16-year-old girl.		
SYMPTOMS:	Menses last 8-10 days, <u>passage of clots</u> , changing <u>protection every 1-2 hours</u> . Occasional nosebleeds.		
HISTORY:	No surgeries, no major trauma.		
VITALS/LABS:	Ferritin: 8 ng/mL		
	Hemoglobin: mildly reduced.		
	PT/aPTT: normal.		VWF Ag:
	VWF Act: 44 IU/dL.		41 IU/dL.
	FVIII: 52 IU/dL.		

DIAGNOSTIC OUTPUT

THE “UNTESTED” TRAP

The absence of surgical bleeding is meaningless; she has never had surgery.

THE MAJOR CHALLENGE

Her uterus has already served as a major hemostatic stress test.

THE HIDDEN METRIC

Ferritin of 8 ng/mL proves chronic mucosal bleeding despite normal PT/aPTT.

CONCLUSION: The normal routine labs and negative surgical history are irrelevant against the overwhelming evidence of the mucosal challenge.

1. Do not ask only “Does the patient bleed?”

Ask what kind of bleeding, in what setting, and after what specific challenge.

2. Treat Heavy Menstrual Bleeding as hematologic evidence.

Ferritin is a crucial gauge of **hemostatic failure**, even when hemoglobin holds.

3. Do not overtrust normal screening tests.

A normal PT, aPTT, or PFA-100 **cannot reliably exclude VWD** if the clinical mucosal pattern is persuasive.

***“Bleeding is not a laboratory value.
It is the clinical result of a hemostatic
system meeting real-world challenges.”***