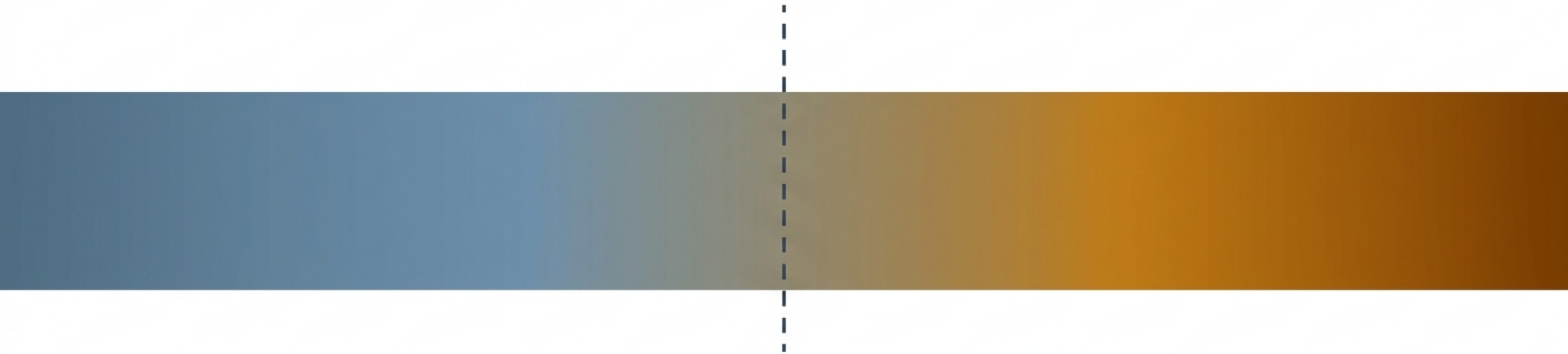
The Borderlands of Diagnosis

Living Between Variation and Disease in von Willebrand Disease



Based on the clinical framework by William Aird

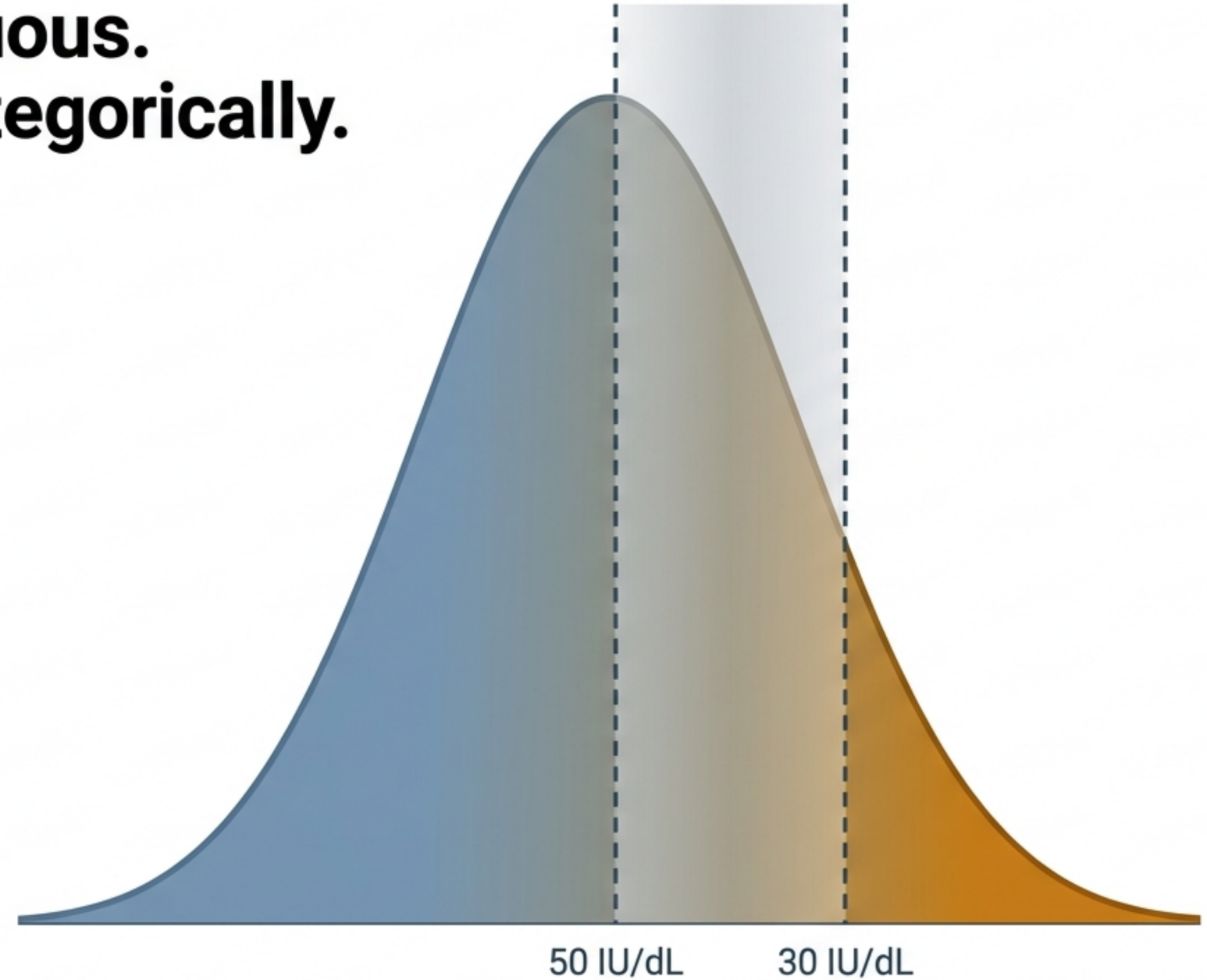
Biology is continuous. Medicine acts categorically.

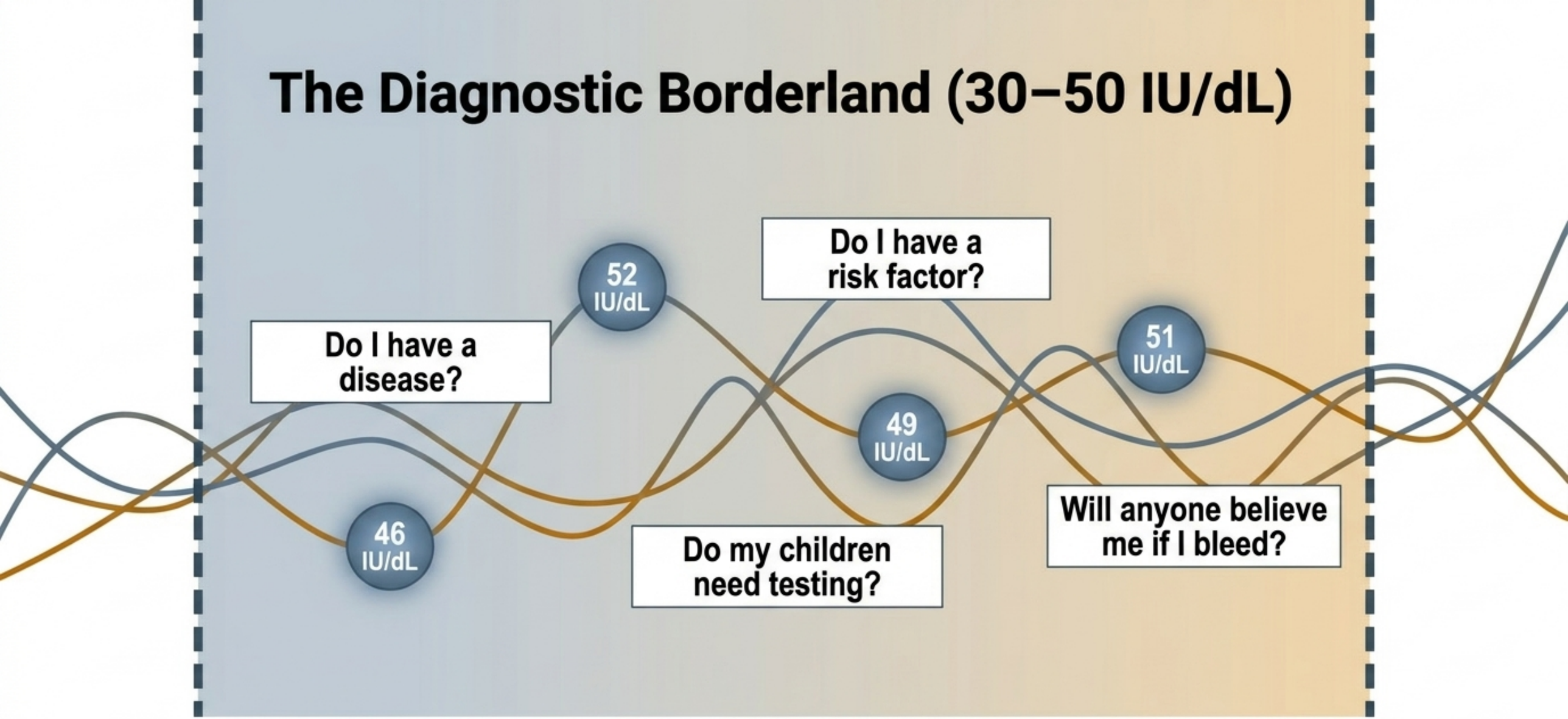
Medicine relies on thresholds to organize care, enable research, and guide clinical communication. But these thresholds are tools, not cliffs in nature.

The Continuum Problem

The distinction between normal and low VWF is partly arbitrary.

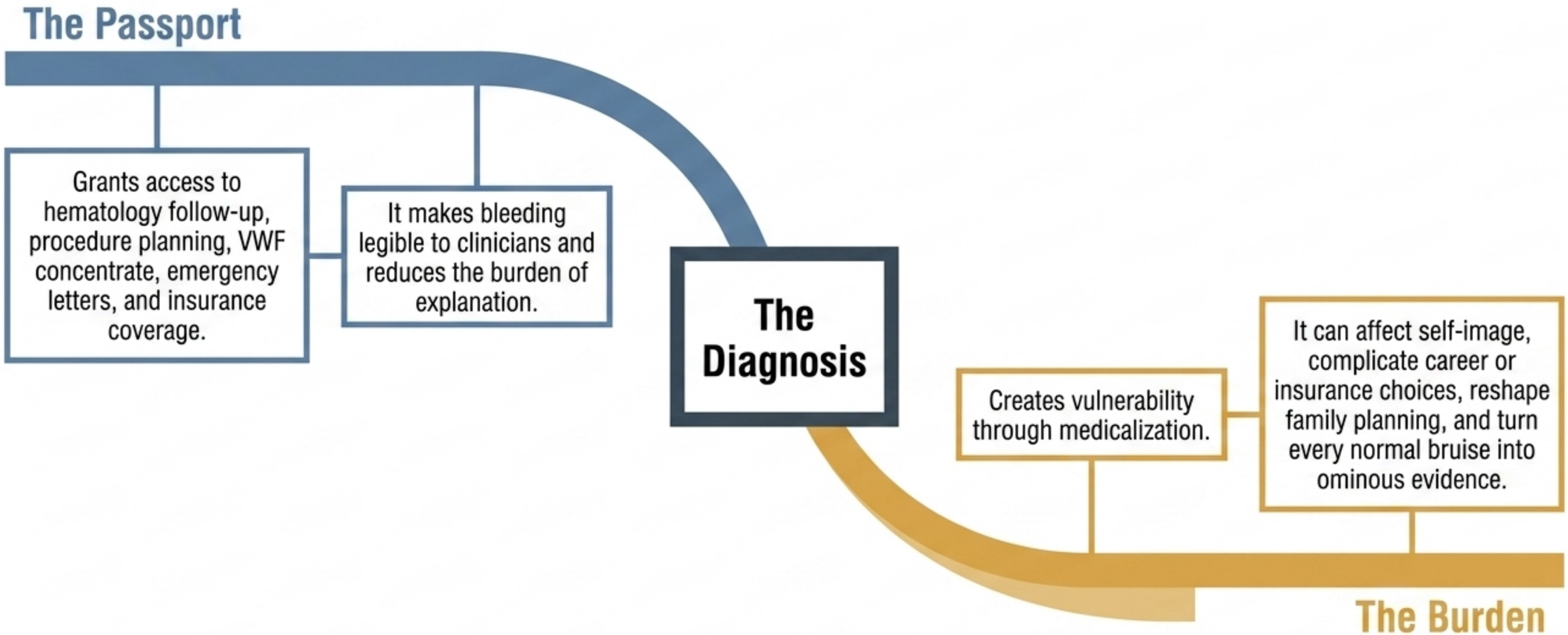
Bleeding risk increases as VWF falls, but the relationship is not strong until VWF is very low.





A patient is just above the line. Or just below it. One clinician says normal. Another says low. Another says type 1 VWD. A value of 49 IU/dL and 51 IU/dL may not represent biologically different states, but clinically, they are treated differently.

The Diagnosis as Passport vs. Burden



The patient near the boundary needs neither reflex labeling nor reflex dismissal. They need careful interpretation.

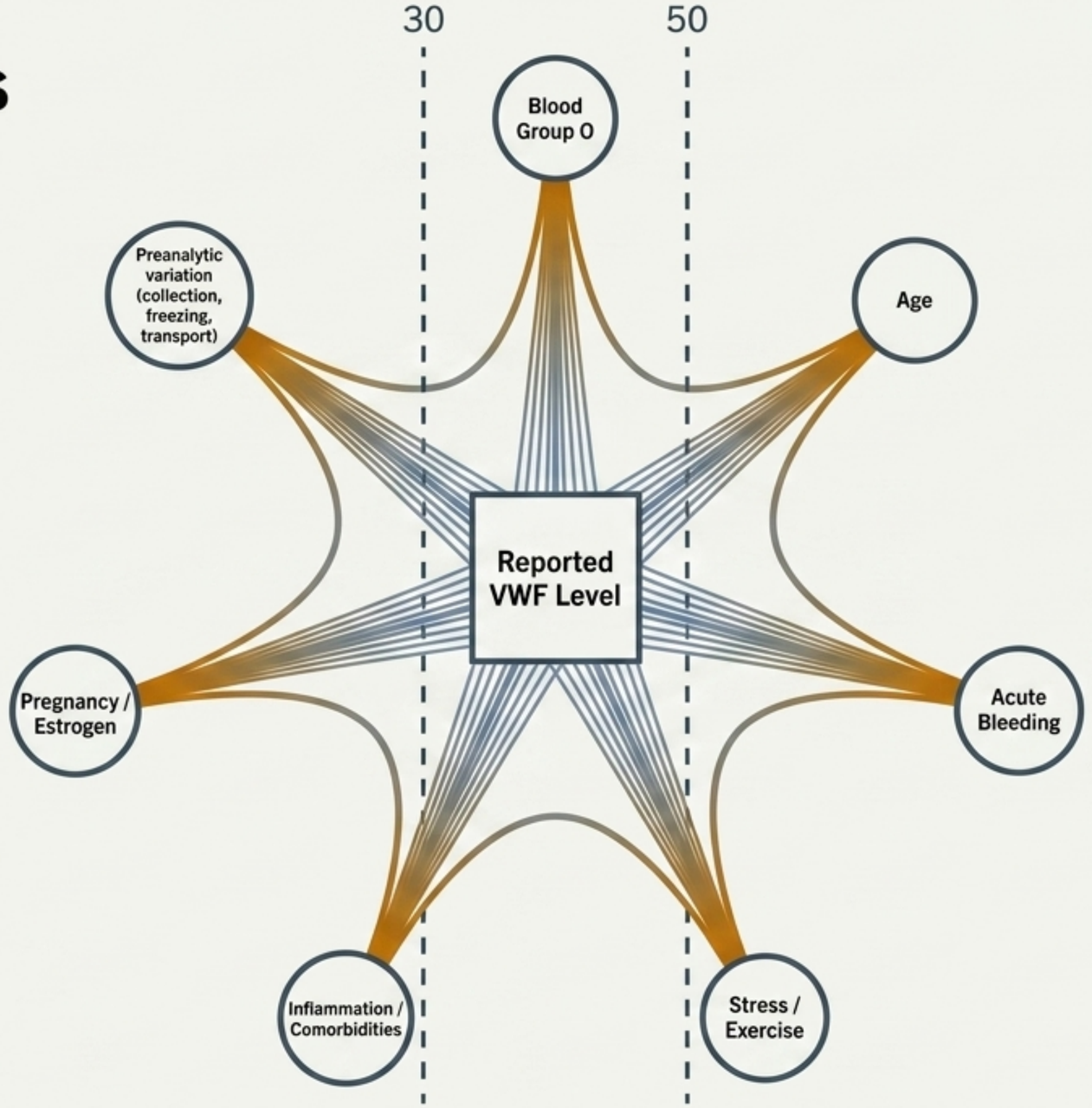
Guidelines Differ Because Tradeoffs Differ

Different guidance documents operationalize different acceptable errors (missed diagnosis vs. **overmedicalization**).

	Older UK/Nordic	2021 ASH/ISTH/NHF/WFH	2024 BSH
Diagnostic Threshold	Cautious approach to 30-50 IU/dL.	Type 1 VWD for <0.30 IU/mL, and <0.50 IU/mL if abnormal bleeding present.	Emphasizes caution in 30-50 IU/dL range.
Approach to 30-50 IU/dL	Treated as a bleeding risk factor rather than definite VWD.	Moves away from “low VWF” VWF” for symptomatic pate.	Notes weaker association between VWF level and bleeding in this range.
Primary Value Protected	Avoiding overdiagnosis and unnecessary labeling.	Avoiding missed diagnosis and ensuring access to care.	Balancing incomplete investigation against the hazards of unnecessary diagnosis.

Assay Uncertainty Becomes Identity Uncertainty

Laboratory numbers feel precise, but VWF testing is vulnerable to context. The patient does not experience an assay artifact; they experience a statement about themselves.



Callout Box

No single test carries the diagnosis. Initial testing requires VWF antigen, platelet-dependent activity, and FVIII. The laboratory contributes a pattern, it does not replace the patient.

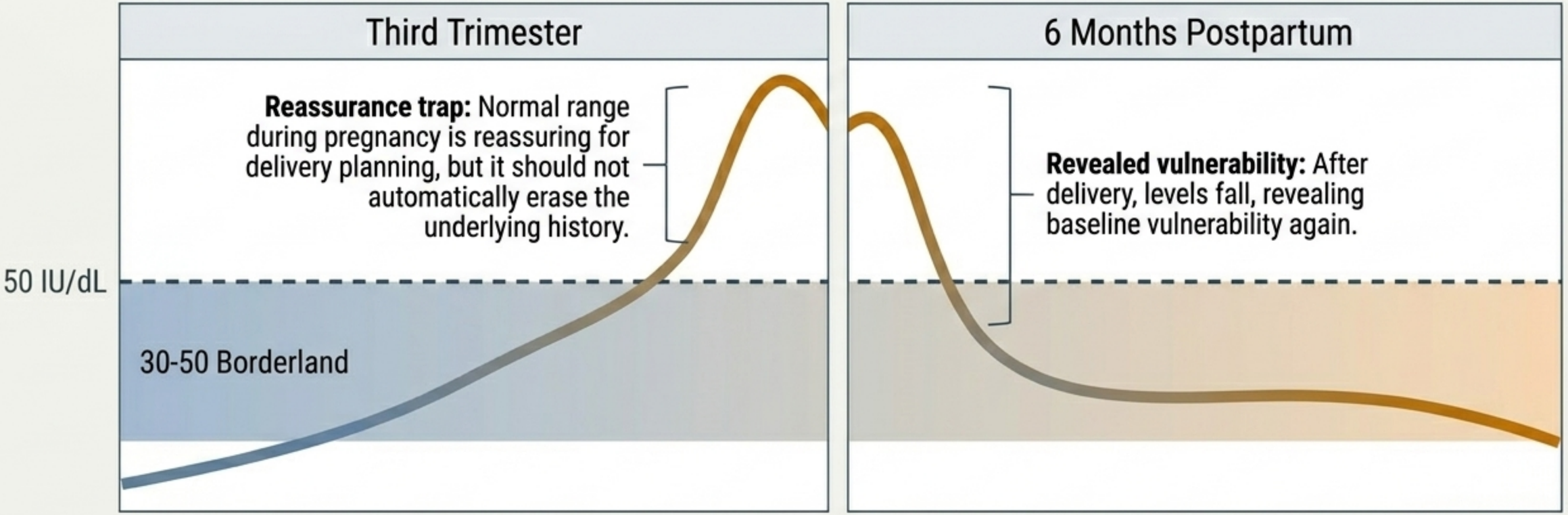
Aging and the Moving Threshold

Aging changes VWF levels. A patient may receive different labels at different stages of life, even if their bleeding phenotype remains similar. The first VWF test is not just a result; it is a timestamp.

Rising levels do not erase historical physiology.

The Pregnancy Borderland

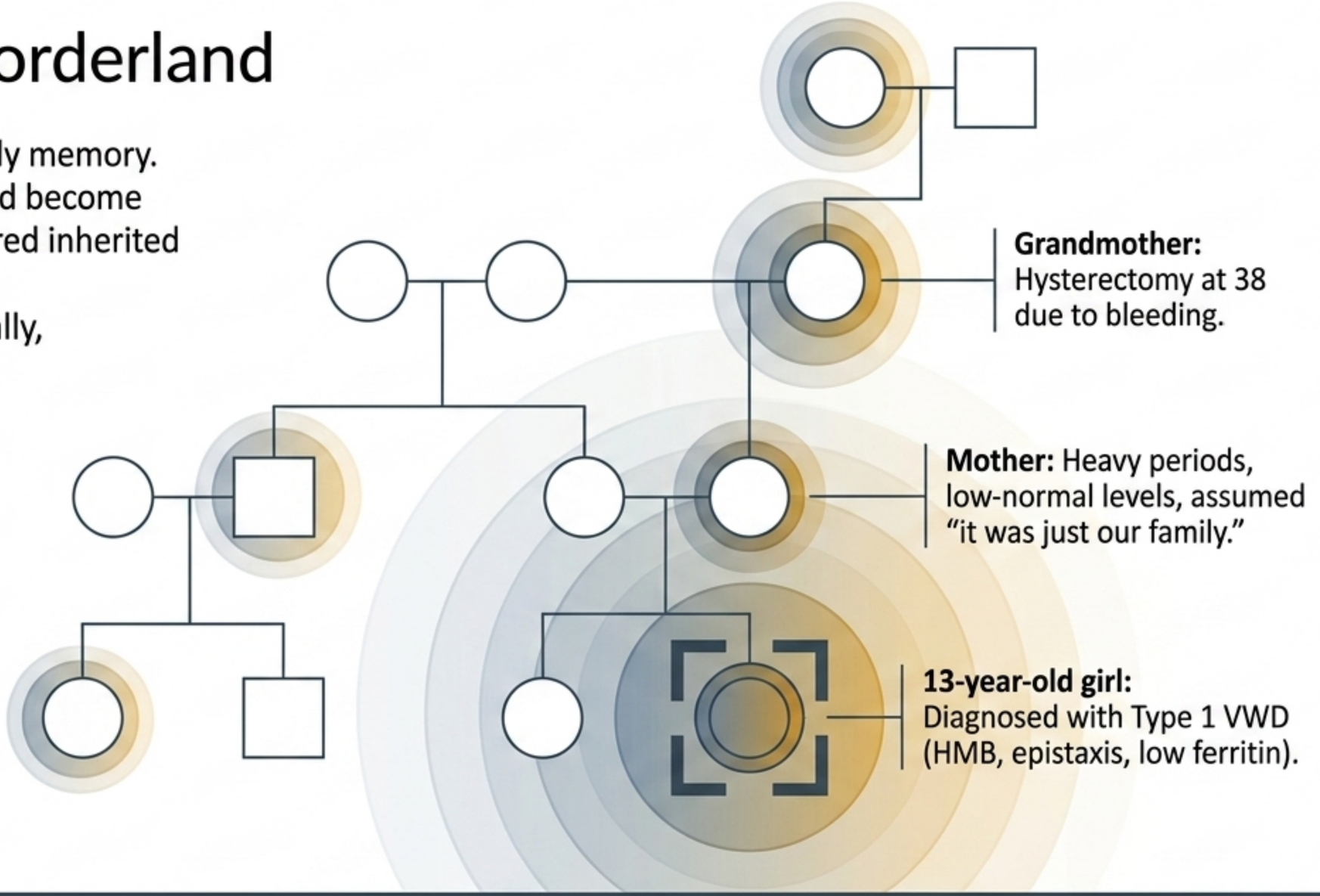
Pregnancy creates temporary physiologic normalization. VWF and FVIII often rise, especially in type 1 VWD and low VWF.



Unexposed does not mean unaffected. The timing of the threshold matters.

The Family Borderland

Diagnosis reorganizes family memory.
Events that seemed isolated become connected as part of a shared inherited pattern.
VWD is inherited biologically, but lived relationally.



Key Takeaway: Inheritance is not fault. Relatives may share the inherited tendency while experiencing different bleeding phenotypes. The goal is anticipatory planning, not creating a family of patients.

The Experience of Being “Undiagnosed”

Removing a diagnosis can reduce anxiety and procedural overplanning.
But thoughtful “undiagnosing” requires more than saying current labs are normal.

The Danger of Erasure

Patients may feel years of bleeding are reinterpreted as insignficant, or worry future clinicians won’t take them seriously.

The Ethics of Revision

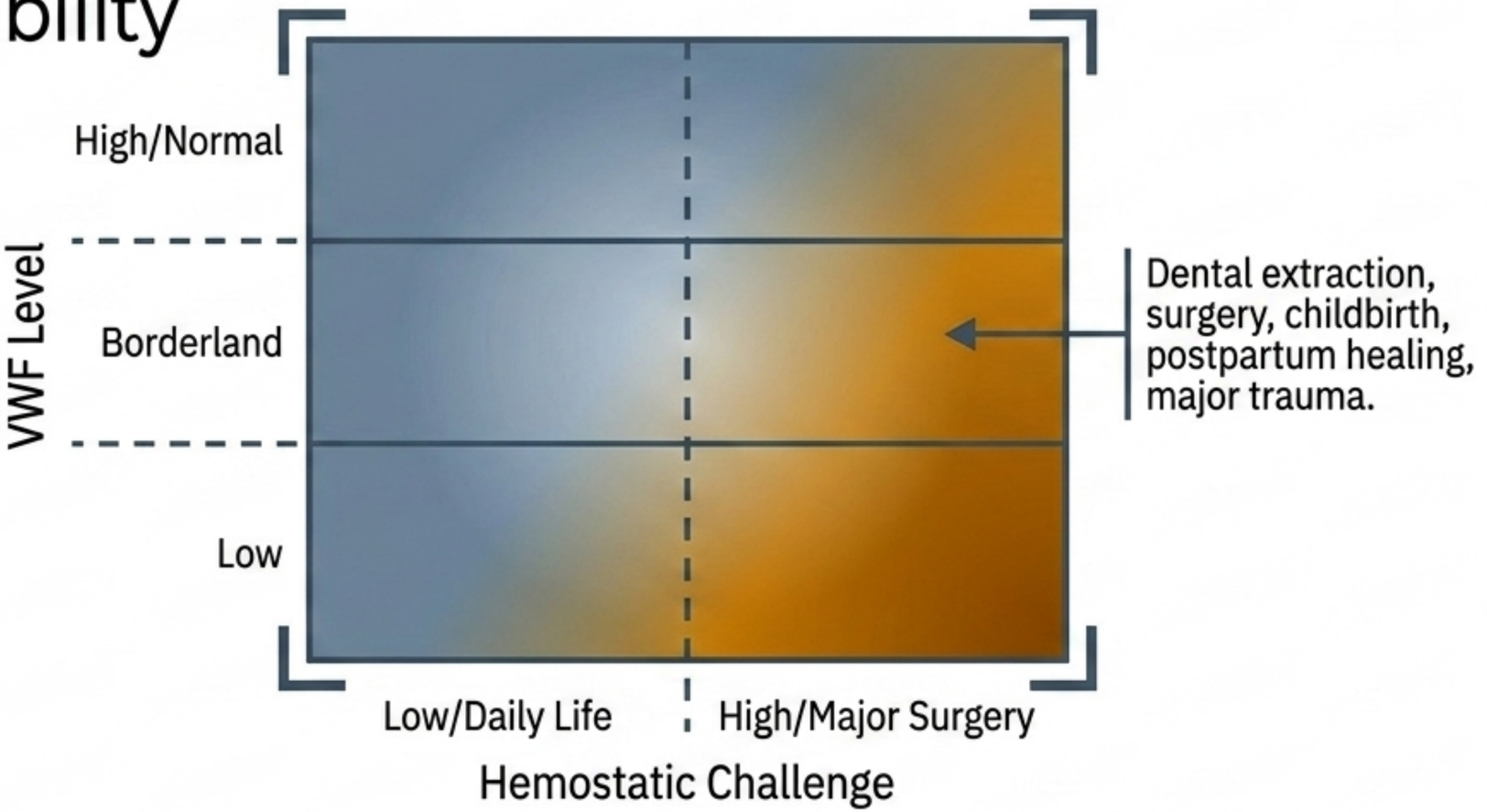
Reassessment should distinguish between current laboratory normalization and the historical question of whether the patient had clinically meaningful risk.

The diagnosis should not be removed by erasing the story.
It should be revised by explaining the story better.

The Paradigm Shift: Conditional Vulnerability

Conditional Vulnerability

VWD is often not a disease of constant impairment. It is a disease of context-dependent risk. The binary of "disease vs. no disease" is too crude near the threshold.



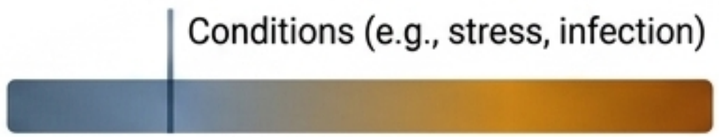
Core Insight: Safety depends more on preparation than on perfect classification.
The practical question is not “What is the perfect label?”, but “What situations require a plan?”

The Documentation Formula

Borderland VWD requires a structured account of risk to prevent the label from doing too much work. Let the story carry the uncertainty.

1. The Data Anchor

Lowest documented VWF levels + conditions under which testing occurred.



2. The Bleeding History

Document bleeding before treatment (dental, menstrual, obstetric, traumatic).

Dental | Menstrual | Obstetric | Traumatic

3. The Contextual Modifiers

Family history + Desmopressin response + Prior effective therapies.

4. The Future Plan

Explicitly define which upcoming situations require a hemostatic plan (regardless of provisional label status).

- **Upcoming Surgery**
- **Childbirth**
- **High-Risk Procedures**
- Major Trauma Protocol

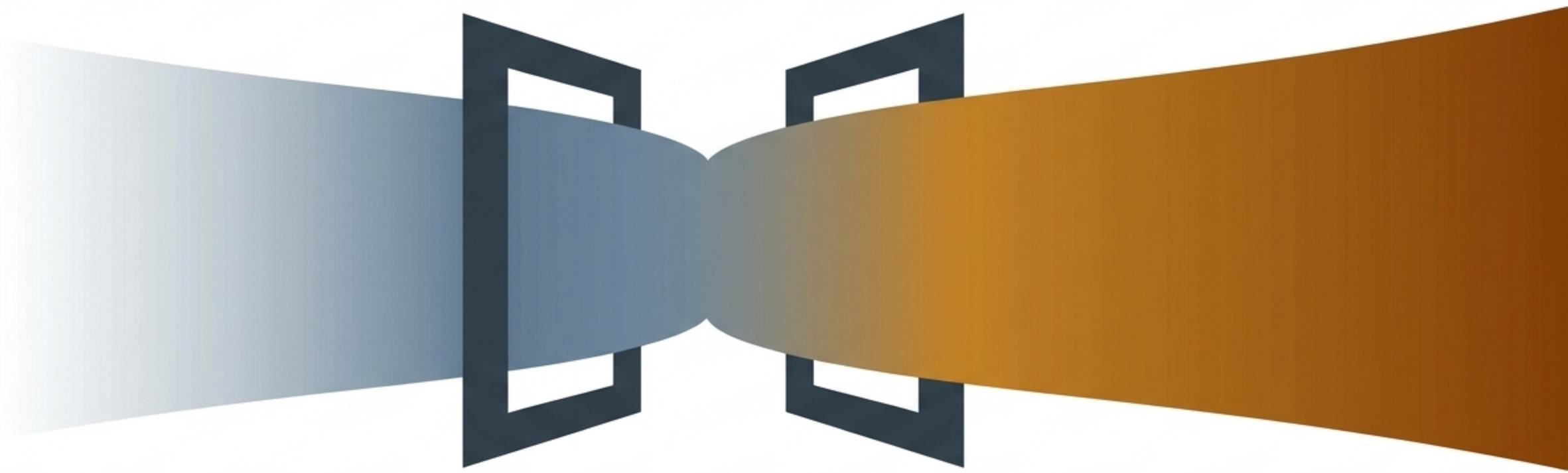
→ **Require Hemostatic Plan**

Naming the Uncertainty: Clinical Language

Instead of (Dismissive/Binary):	Try (Nuanced/Borderland-Aware):
Your labs are normal.	This value is reassuring, but we should interpret it with your prior values, bleeding history, and the situation we are planning for.
You do not quite meet criteria.	You are near the diagnostic boundary. That means we should be careful about both overlabeling and underplanning.
It is probably just blood group O.	Blood group O may contribute to lower VWF levels, but it does not erase your bleeding history.
Your VWF normalized, so you no longer have a bleeding disorder.	Your current VWF level is higher than before. We should reconsider the diagnosis thoughtfully rather than erase the history.

The Threshold is the Beginning

Thresholds are ethically necessary tools for decision-making, not measures of human worth or suffering. In
In the diagnostic borderlands of VWD, the goal is not to pretend the lines are certain, but to make the uncertainty safe.



1. Preserve the bleeding story.

2. Plan for meaningful risk.

3. Speak honestly about uncertainty.

Patients near the line need more than a number. They need an explanation.