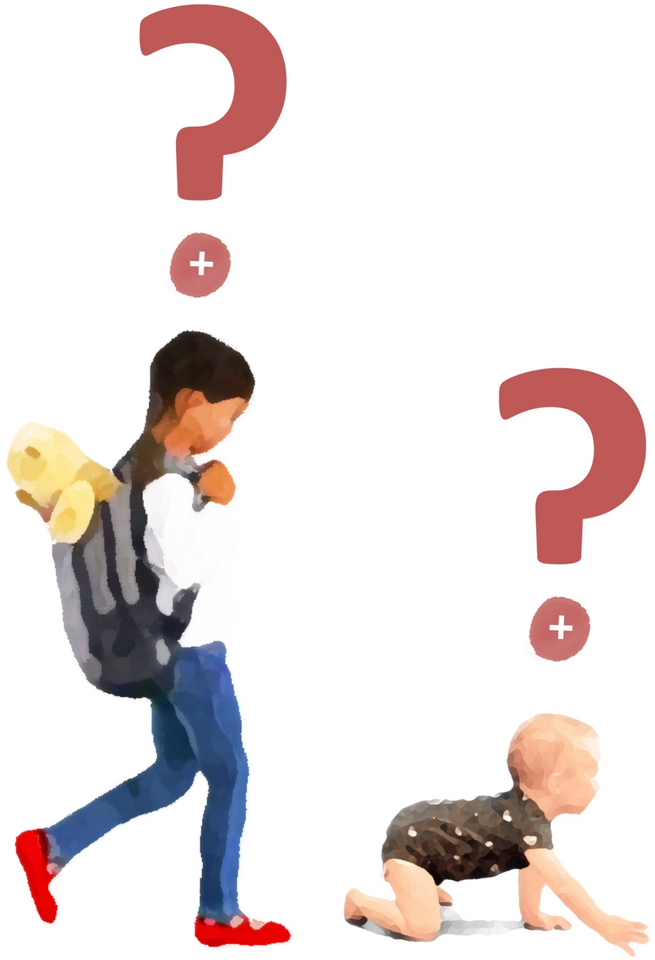
VON WILLEBRAND DISEASE

Module 1

When to consider von Willebrand disease?



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CME credit available
Visit programme home page on:

What will you learn in this micro e-learning programme about von Willebrand disease?

This micro e-learning programme consists of **three modules** aiming to increase awareness, knowledge and understanding of the existence, diagnosis, treatment and implications of von Willebrand disease among healthcare professionals outside of haematology.

Upon completion of the three modules, you will:

- be aware of the potential **existence of von Willebrand disease** in your patient population
- be able to **recognise the signs and symptoms** of von Willebrand disease
- understand **how to search for von Willebrand disease**, effectively using the screening tools available and signposting patients towards earlier diagnosis
- be aware of the **impact** of von Willebrand disease on other care

AFTER MODULE 1 YOU WILL

Current Module

- be aware of the potential **existence of von Willebrand disease** among your patient population
- be able to recognise the **signs and symptoms** of von Willebrand disease

AFTER MODULE 2 YOU WILL

- understand you can **help diagnose** von Willebrand disease
- be able to effectively use the **screening tools** available
- understand the **next steps** to take when suspecting von Willebrand disease

AFTER MODULE 3 YOU WILL

- be aware of the **impact of von Willebrand disease on provision of general care**
- understand the key **aspects of von Willebrand disease affecting your clinical practice**
- understand the importance of a **multidisciplinary approach** in von Willebrand disease care

This micro e-learning module has been developed by a **multidisciplinary panel of experts**

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The views expressed in this slide deck are the personal opinions of the experts.
They do not necessarily represent the views of the experts' institutions or the rest of the HEMOSTASIS CONNECT group.

Von Willebrand disease
at a glance

Von Willebrand disease is the **most common inherited bleeding disorder.**[1]

- It is estimated to occur in 1 in 100-1000 people, although tertiary care referrals for von Willebrand disease occur at a much lower frequency (˜1 in 10,000)
- von Willebrand disease occurs in men, women, and children

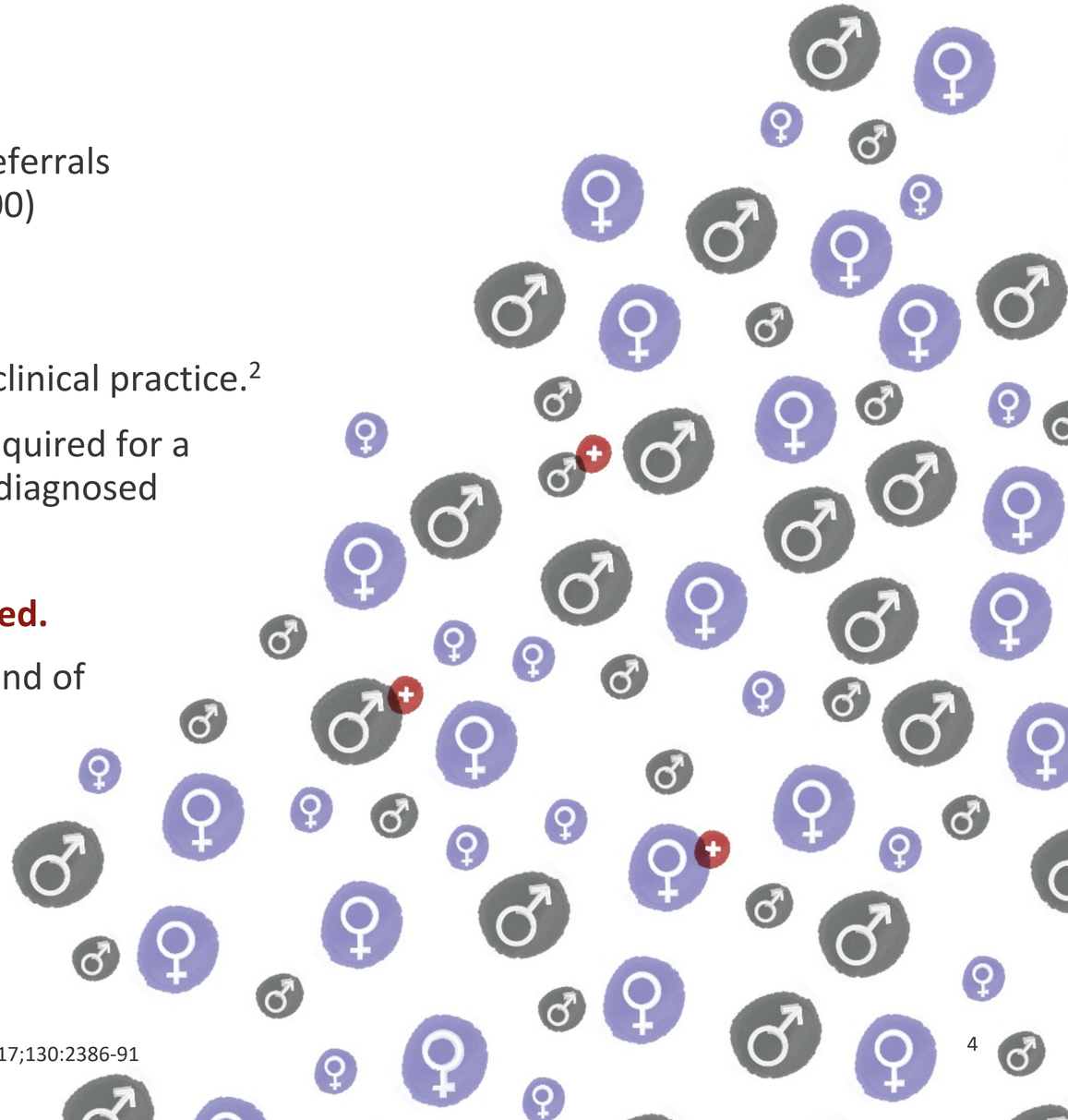
Von Willebrand disease is one of the most **overlooked** entities in everyday clinical practice.[2]

- The complexity and heterogeneity of the disease and breadth of tests required for a formal diagnosis lead it to be under-diagnosed, over-diagnosed and misdiagnosed

Even if the diagnosis is made, von Willebrand disease is often **underestimated.**

- It has a significant impact on quality of life and on the provision of any kind of other healthcare provided to the patient[3]

! It is important not to miss von Willebrand disease

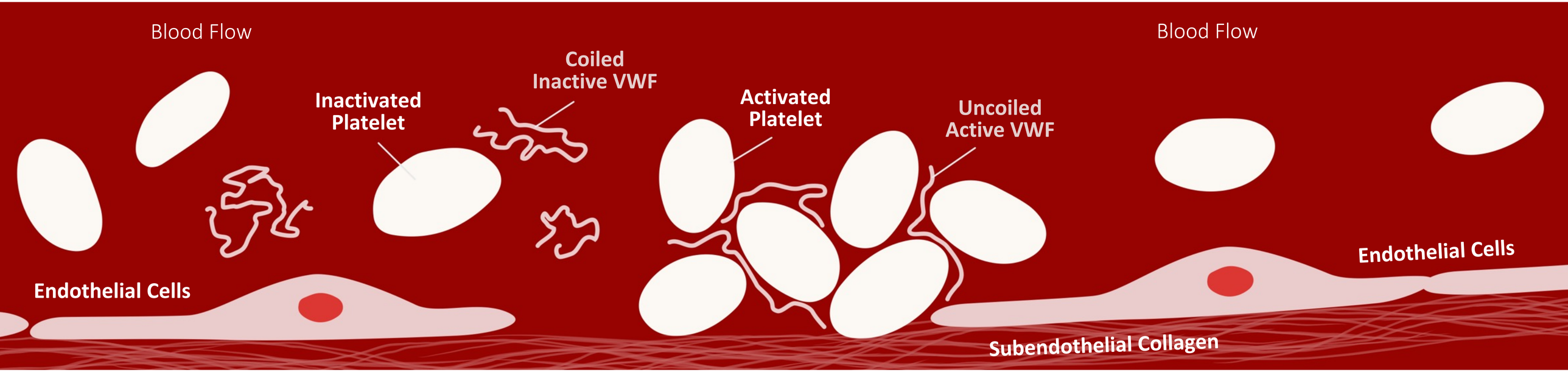


1. Bowman M, et al. J Thromb Haemost. 2010;8:213-6; 2. Colonne CK, et al. J Blood Med. 2021;12:755-68; 3. Sharma R, et al. Blood. 2017;130:2386-91

Von Willebrand disease is a **disorder of primary haemostasis**

- **Von Willebrand factor (VWF)** binds to collagen at sites of vascular injury and **mediates platelet adhesion and aggregation**
- Patients with von Willebrand disease have reduced levels of (functional) VWF, leading to defective platelet adhesion and aggregation
- In patients with severe von Willebrand disease, secondary haemostasis can also be affected, as VWF is a carrier of Factor VIII
- There are various types of von Willebrand disease
- The **multifunctional nature of VWF** explains the heterogeneity in clinical symptoms and bleeding risk of von Willebrand disease, as well as the diagnostic challenges

Type 1	**Quantitative disorder of VWF**
Type 2	**Qualitative and quantitative disorder of VWF** • Various subtypes
Type 3	**Absolute lack of VWF**

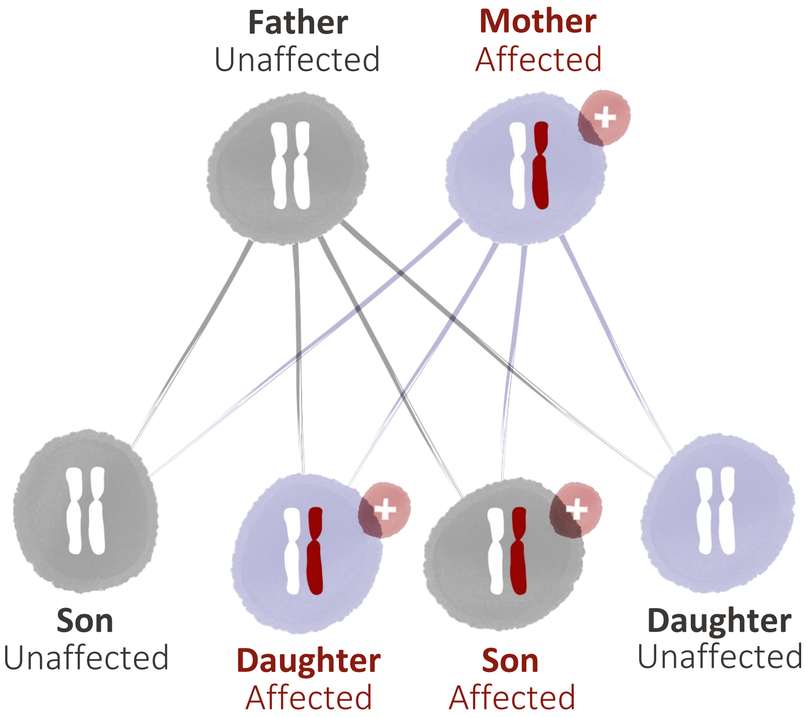


VWF, von Willebrand factor

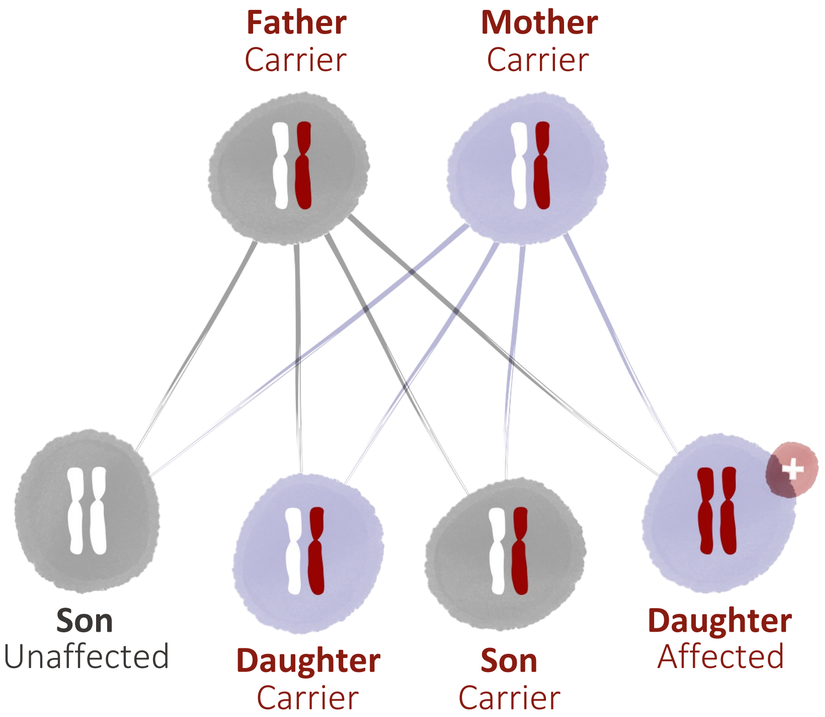
Colonne CK, et al. J Blood Med. 2021;12:755-68; Leebeek FWG, Eikenboom JCJ. N Engl J Med. 2016;375:2067-80

Von Willebrand disease is a **hereditary disease** that occurs in men and women

von Willebrand disease typically has an **AUTOSOMAL DOMINANT** pattern of inheritance



Type 3 von Willebrand disease typically has an **AUTOSOMAL RECESSIVE** pattern of inheritance



Although a genetic mutation is not commonly found in mild cases of von Willebrand disease, it is still a familial disorder
In rare cases, von Willebrand disease can be acquired later in life

Leebeek FWG, Eikenboom JCJ. N Engl J Med. 2016;375:2067-80; Eikenboom JC. Best Pract Res Clin Haematol. 2001;14:365-79; Mital A. Adv Clin Exp Med. 016;25:1337-44

Signs and symptoms von Willebrand disease

General

- **Family history** of a bleeding disorder
- Notable **bruising** without injury
- **Prolonged/excessive bleeding**, even from minor wounds

Dental

- Prolonged bleeding following invasive **dental procedures**
- Prolonged **bleeding from the gums** following deep cleaning
- **Recurrent ulcers and pallor** of the mucosa associated with anaemia

Paediatric

- **Nosebleeds**
- Bleeding **during teething** in small children
- Notable **bruising without injury**
- **Excessive bleeding** from minor wounds

ENT

- **Nosebleeds**
- Severe (potentially life-threatening) bleeding after **tonsillectomy/adenoidectomy**

Gynaecological

- Heavy **menstrual bleeding**, especially since menarche
- Bleeding during **ovulation**
- Primary and late **post-partum haemorrhage**

Surgical

- **Joint pain** and/or bleeds
- Prolonged and/or severe bleeding after **minor or major surgery**

Gastrointestinal

- **Bleeding of gastrointestinal tract** with or without an obvious anatomic lesion in adults

Von Willebrand disease is mainly a disorder of primary haemostasis, so the bleeding starts quickly, not delayed like in haemophilia

ENT, ear, nose, and throat
This slide is based on the clinical experience of the authors

Next steps
when suspecting von Willebrand disease

It is not easy to diagnose von Willebrand disease, as there is a large variability in symptoms and interpretation of laboratory tests is complex.

When suspecting von Willebrand disease, **refer the patient to a haematologist** for diagnostic laboratory assessment.

Please refer to the **other modules of this micro e-learning programme** for more information on:

- **Module 2:** Screening and next steps
- **Module 3:** A multidisciplinary approach: impact of von Willebrand disease on the provision of general care

Summary

von Willebrand disease is the **most common inherited bleeding disorder,** occurring in men, women and children

Signs and symptoms of von Willebrand disease include easy bruising, heavy menstrual bleeding, nosebleeds and prolonged bleeding after surgery

When suspecting von Willebrand disease, **refer to a haematologist** for diagnostic laboratory assessment

Next steps

Please now proceed to the **assessment quiz** in the e-learning to test your knowledge.

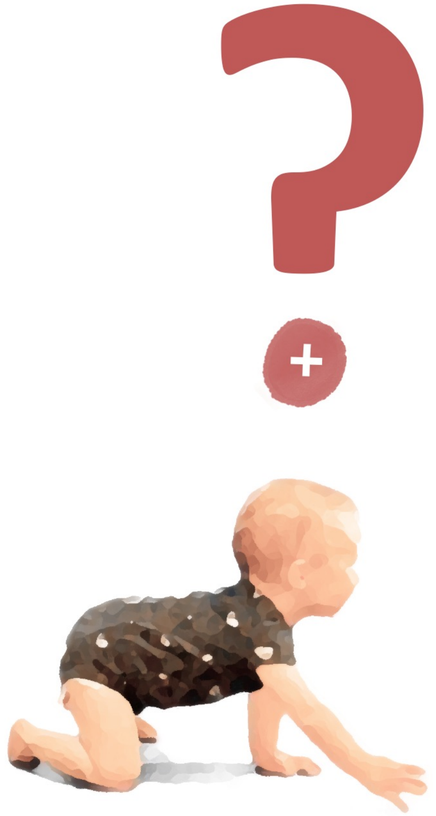
Visit **Module 2** of this micro e-learning programme to learn more about:

- how you can help diagnose von Willebrand disease
- how to effectively use the screening tools available
- the next steps to take when suspecting von Willebrand disease

Visit **Module 3** of this micro e-learning programme to learn more about:

- the impact of von Willebrand disease on provision of general care
- the key aspects of von Willebrand disease affecting your clinical practice
- the importance of a multidisciplinary approach

Note: you will be able to claim your CME credit after passing at least 2 of the 3 modules



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