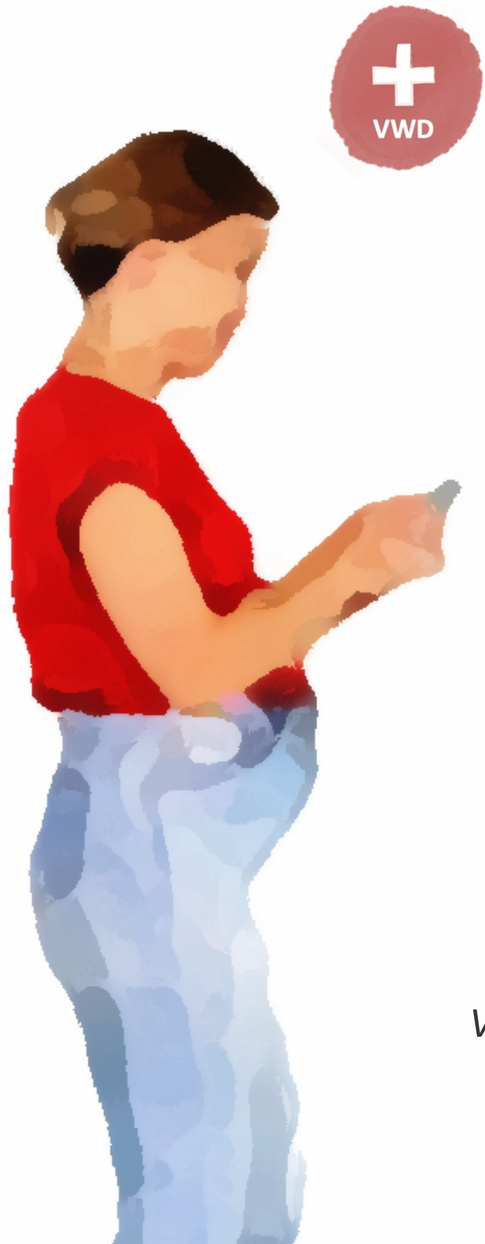
HEMOSTASIS connect
POWERED BY COR2ED

VON WILLEBRAND DISEASE

Module 3

How to care for a patient with von Willebrand disease?



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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

- 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

Current Module

- 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

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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.

When to suspect von Willebrand disease
Module 1 summary

Module 1 of this micro e-learning programme described how von Willebrand disease is the most common inherited bleeding disorder, occurring in men, women, and children. **Signs and symptoms** of von Willebrand disease include:

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**
- **Prolonged/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

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

When to suspect von Willebrand disease
Module 2 summary

Module 2 of this micro e-learning programme described the **next steps** to take when you **suspect von Willebrand disease** in your patient:

1

History

Take a **thorough bleeding history,** including a detailed family history

- Use a validated **bleeding assessment tool (BAT),** such as the ISTH-SSC BAT

2

Laboratory assessment

Perform general laboratory assessment

- Assessment of iron status, haemoglobin, and red blood cell count provide important information for clinical management

Refer to/consult a haematologist for diagnostic laboratory assessment

- There are many pitfalls in the interpretation of diagnostic laboratory tests

3

Management

Management of von Willebrand disease requires a **multidisciplinary approach**

Surgical procedures require a **multidisciplinary risk assessment** and proportionate **personalised management plan**

BAT, bleeding assessment tool; ISTH, International Society on Thrombosis and Haemostasis; SSC, Scientific and Standardization Committee

Treatment of von Willebrand disease

Several treatment options are available to **improve haemostasis**

- They are used to **manage** symptoms and **prevent** excessive/prolonged bleeding after surgery or delivery

The **treatment choice** is impacted by:

- Type and severity of von Willebrand disease
- Clinical circumstances

Oral or intravenous anti-fibrinolytics *(tranexamic acid or aminocaproic acid)*
bind to the lysine-binding sites of plasminogen and delay the breakdown of blood clots

Nasal or intravenous DDAVP *(desmopressin)*
induces release of VWF from storage sites in endothelial cells

(Fluid restrictions are required)

Intravenous VWF replacement therapy
directly supplements (functional) VWF

Other commonly used treatment options in patients with von Willebrand disease:

- Hormonal therapy for the treatment of heavy menstrual bleeding
- Iron replacement therapy for the treatment of iron deficiency with or without anaemia

DDAVP, desmopressin; VWF, von Willebrand factor
Connell NT, et al. Blood Adv. 2021;5:301-25

Von Willebrand disease
impacts many aspects of healthcare

All healthcare practitioners should be **aware of the diagnosis** of von Willebrand disease and **its impact on the care they provide.**

Psychological and primary care

- Quality of life issues
- Misinterpretation of bruising

Surgery

- Prolonged/excessive bleeding during or after surgery
- Orthopaedic surgery for arthropathy (rare*)

Anaesthesia and pain management

- Neuraxial anaesthesia
- NSAID use
- Increased risk for needing a blood transfusion

Dentistry

- Prolonged/excessive bleeding during or after invasive dental procedures

Obstetrics and gynaecology

- Heavy menstrual bleeding
- Prolonged/excessive bleeding with pregnancy, delivery, and fertility procedures

Invasive procedures

- Prolonged/excessive bleeding during or after invasive procedures, such as endoscopy or fertility procedures

Multidisciplinary care is key, building of **network of teams** who provide wider care for patients with von Willebrand disease, including nurses, dental care specialists, surgeons, primary care physicians, psychologists, etc.

In many countries, diagnosed patients receive an **emergency card** from their haematologist to show to healthcare providers in case of emergency, **helping patients to advocate for themselves.**

* von Willebrand disease is rarely associated with arthropathy requiring orthopaedic surgery; this complication is more common with haemophilia
NSAID, non-steroidal anti-inflammatory drug

Careful planning
is required with invasive procedures, pregnancy and delivery

On a day-to-day basis, von Willebrand disease may not cause major issues.

However, there are specific circumstances where **caution** should be exercised, such as:

- **Pregnancy**
- **Delivery**
- **Minor elective surgery**
- **Major surgery**
- **Invasive procedures (e.g., endoscopy, fertility procedures)**

Patients with von Willebrand disease have an increased bleeding risk, so **involve a haematologist early, as careful planning is required.**

- Ensure a **haemostasis care plan** is in place for the prevention and treatment of bleeding complications

Do not hesitate to contact the haematologist, as it is preferable to **avoid bleeding, instead of treating it** when it happens.

- It is important not to **under-treat** patients with von Willebrand disease

Planning care
for a patient with von Willebrand disease

Consult a haematologist upfront whenever there is concern for bleeding risk

To decide on the most appropriate haemostasis care plan, the haematologist will consider:

Type and severity of von Willebrand disease

Patients with **severe disease** may need VWF replacement therapy, where in patients with **milder phenotypes** anti-fibrinolytics or DDAVP may suffice

In **milder** von Willebrand disease, **pregnancy and surgery can increase VWF levels** (sometimes to normal range), but for others additional therapy may be needed

Bleeding risk associated with the situation or procedure

Pregnant patients should be followed up in a **high-risk antenatal clinic**

If VWF levels do not sufficiently increase during pregnancy, additional treatment is required for delivery

VWF levels drop significantly after delivery, so awareness of **postpartum bleeding** is required

For **minor procedures**, antifibrinolytics may be sufficient to manage the bleeding risk

Other circumstances (such as comorbidity)

DDAVP may not be suitable for some patients, including elderly patients with an increased cardiovascular risk and patients who cannot tolerate the fluid restrictions

DDAVP, desmopressin; VWF, von Willebrand factor
Doherty D, et al. J Thromb Haemost. 2021;19:701-10

Planning dental care
for a patient with von Willebrand disease

Dental Care

Gum bleeding is common but often ignored in patients with VWD.[1-3]

Healthcare providers and patients mistakenly think gum bleeding is acceptable or inevitable and caused by the VWD itself from brushing too hard. Untreated, this leads to periodontal disease and tooth loss over time.

Managing the risk

- Educate patients on how to care for their gums
- Give patients permission to brush in the presence of bleeding to ensure the plaque biofilm is removed thoroughly to reduce local inflammation

Dental procedures

Dental cleaning, fillings, sealants and dental injections are usually not high-risk procedures.

Higher-risk procedures include gum surgery or dental extractions.

Managing the risk

- Dental cleaning can usually be carried out under anti-fibrinolytic cover alone[4,5]
- Dental procedures require upfront risk assessment and careful planning
- Higher-risk procedures require a dentist applied local measures, systemic antifibrinolytic therapy and/or replacement therapy plus careful post-operative instructions

VWD, von Willebrand disease

1. Carcao MD, et al. Haemophilia. 2010;16:943-8; 2. Noone D, et al. Haemophilia. 2021;27 issue S2:143; 3. Epping L, et al. PLoS One. 2018;13:e0191291;
4. Doherty D, et al. J Thromb Haemost. 2021;19:701-10; 5. van Galen KP, et al. Cochrane Database Syst Rev. 2019;4:CD011385

Planning care for a patient with von Willebrand disease: **the role of the patient**

Before surgery, patients should:

- **Know and be able to summarise** their type of von Willebrand disease, bleeding history and prior treatments
- Be aware of any **pre-operative preparation** (e.g., starting oral anti-fibrinolytic therapy)
- Optimise their **nutritional and iron status**
- **Understand the procedure and the bleeding risk**
- Be aware of their **options for pain relief**
  - Note NSAIDs are relatively contraindicated

Before higher-risk dental procedures, patients should:

- **Time** the dental appointment to gain optimal protection from the systemic haemostatic measures and **plan** procedures at the beginning of the week, so any post-procedural bleeding occurs during weekdays, not on the weekend
- **Thoroughly clean the teeth** during the week before, to remove plaque and reduce local inflammation that may increase bleeding from the wound
- Shop for **soft foods** to prevent traumatising the wound and causing a rebleed
- Prepare to **not smoke** post-operatively; nicotine replacement patches may be of help
- Start **oral antifibrinolytics** the evening before or the morning of the procedure (if applicable)
- **Do not leave the dental clinic until haemostasis has been gained** using local measures, including biting on s-gauze pack for at least 20-30 minutes

NSAID, non-steroidal anti-inflammatory drug
This slide is based on the clinical experience of the authors.

Acute care
for a patient with von Willebrand disease

In case of acute medical situations…

Bleeding events or other conditions requiring acute medical care

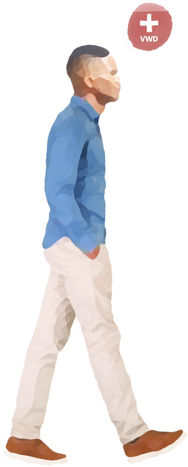
Prepare for excessive bleeding and prioritise the bleeding

Ask the patient for the type of von Willebrand disease, bleeding history and treatment of choice

(or check the guidelines)

If possible, always consult a haematologist

Inform the primary care physician where applicable



The role of the patient in acute care

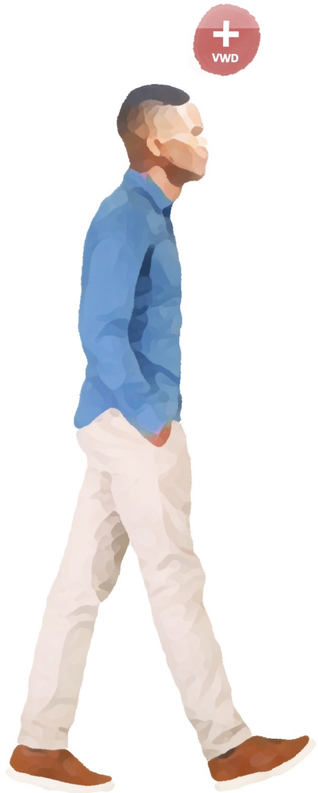
Make sure:

- patients know the local emergency pathway and emergency contact numbers
- any factor replacement patients have at home is always in date *(if applicable)*
- when travelling, patients have international insurance and are aware of the nearest haemophilia centres *(for patients with a more severe bleeding phenotype), as well as have a supply of any (emergency) treatment they may need (e.g. tranexamic acid, von Willebrand factor)*

Caring for a patient with von Willebrand disease

Von Willebrand disease is a chronic disorder, so the key is to **support patients in living a 'normal' life.**

Allow time for patients to discuss issues relating to the impact of the bleeding disorder on their daily life.



Psychological/ practical support

Work
School
Relationships
Sport
Manual jobs
Travel
Recurrent nosebleeds
Heavy menstrual bleeding
Emergency help

Lifestyle support

Exercise
Diet
Weight
Iron supplements

Von Willebrand disease may influence **concomitant medication.**

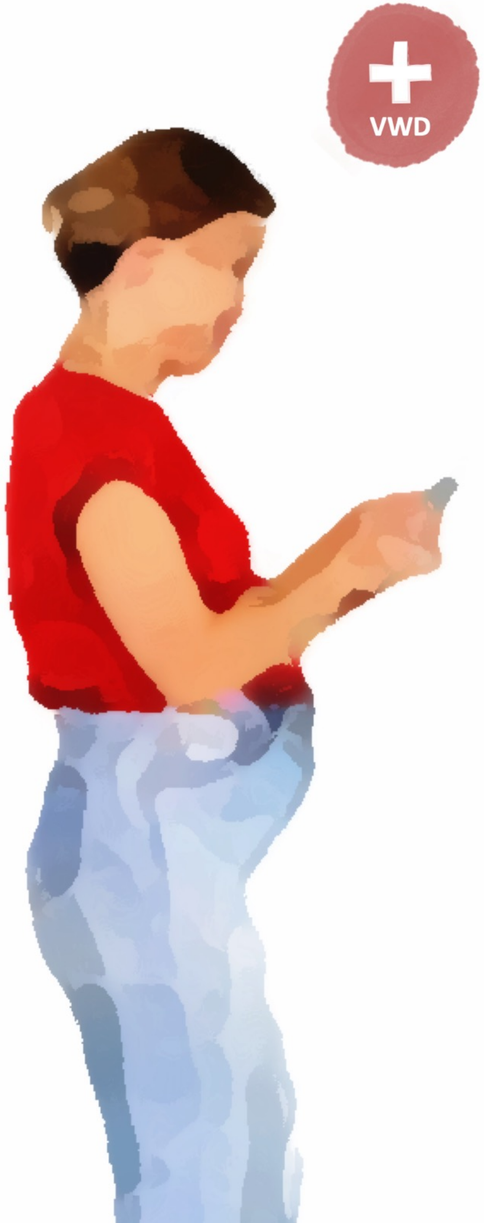
- **NSAIDs and aspirin** are relatively contraindicated, as they influence platelet function[1]
- Paracetamol and COX-2 inhibitors can be used for pain relief
- If NSAIDs or aspirin are needed, carefully consider risks and benefits on a case-by-case basis
- Before starting any **new medication, check for a potential impact on clotting** and discuss the individual risk-benefit profile with a haematologist

Patient education

- Use written information to reinforce support and aid decision making
- Patient advocacy groups often provide helpful information and support

Consider **at least yearly haematology follow up.**

Summary



Von Willebrand disease impacts many aspects of healthcare, including planned and acute care, so a **multidisciplinary approach** is required

Consult a haematologist upfront whenever there's concern for bleeding risk, to develop a haemostatic treatment plan and be actively involved in the care team

Von Willebrand disease may **influence the choice of other medication** patients may need, such as pain medication

Next steps

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

Visit **Module 1** of this micro e-learning programme to learn more about:

- the potential existence of von Willebrand disease among your patent population
- the signs and symptoms of von Willebrand disease

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

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

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