

WOMEN WITH
BLEEDING DISORDERS
Module 1

How do I recognise and screen for bleeding disorders in women and girls

March 2021

The programme is supported by an independent educational grant from Takeda.

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What will you learn in this micro e-learning programme on women with bleeding disorders?

This micro e-learning programme consists of **two modules** aiming to increase awareness, knowledge, and understanding of bleeding disorders and their management by healthcare professionals who are the first to see women and girls with signs and symptoms of bleeding disorders.

Upon completion of the two modules, you will:

- Be able to **recognise the signs and symptoms** of bleeding disorders in women and be aware of their **impact on the quality of life** of women
- Know how to **identify women who are likely to have undiagnosed bleeding disorders**, initiate basic diagnostic testing, and arrange an appropriate referral for diagnosis
- Be able to **initiate management** of bleeding symptoms in women until specialist assessment is available

Current Module

AFTER MODULE 1 YOU WILL:

- Be able to **recognise the signs and symptoms** of bleeding disorders in women
- Be aware of the **impact on quality of life** of bleeding disorders in women
- **Know how to effectively screen for symptoms** of bleeding disorders in women by using the tools available

AFTER MODULE 2 YOU WILL:

- **Understand the strengths and limitations of tests** used to diagnose bleeding disorders in women
- Be able to perform **basic interpretation of test results** to diagnose bleeding disorders in women
- Be able to **initiate management** of bleeding symptoms in women until specialist assessment is available

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



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- Chair of the European Association for Haemophilia and Allied Disorders (EAHAD) Women and Bleeding Disorders Committee
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- Associate Professor of Paediatrics at the Ohio State University College of Medicine
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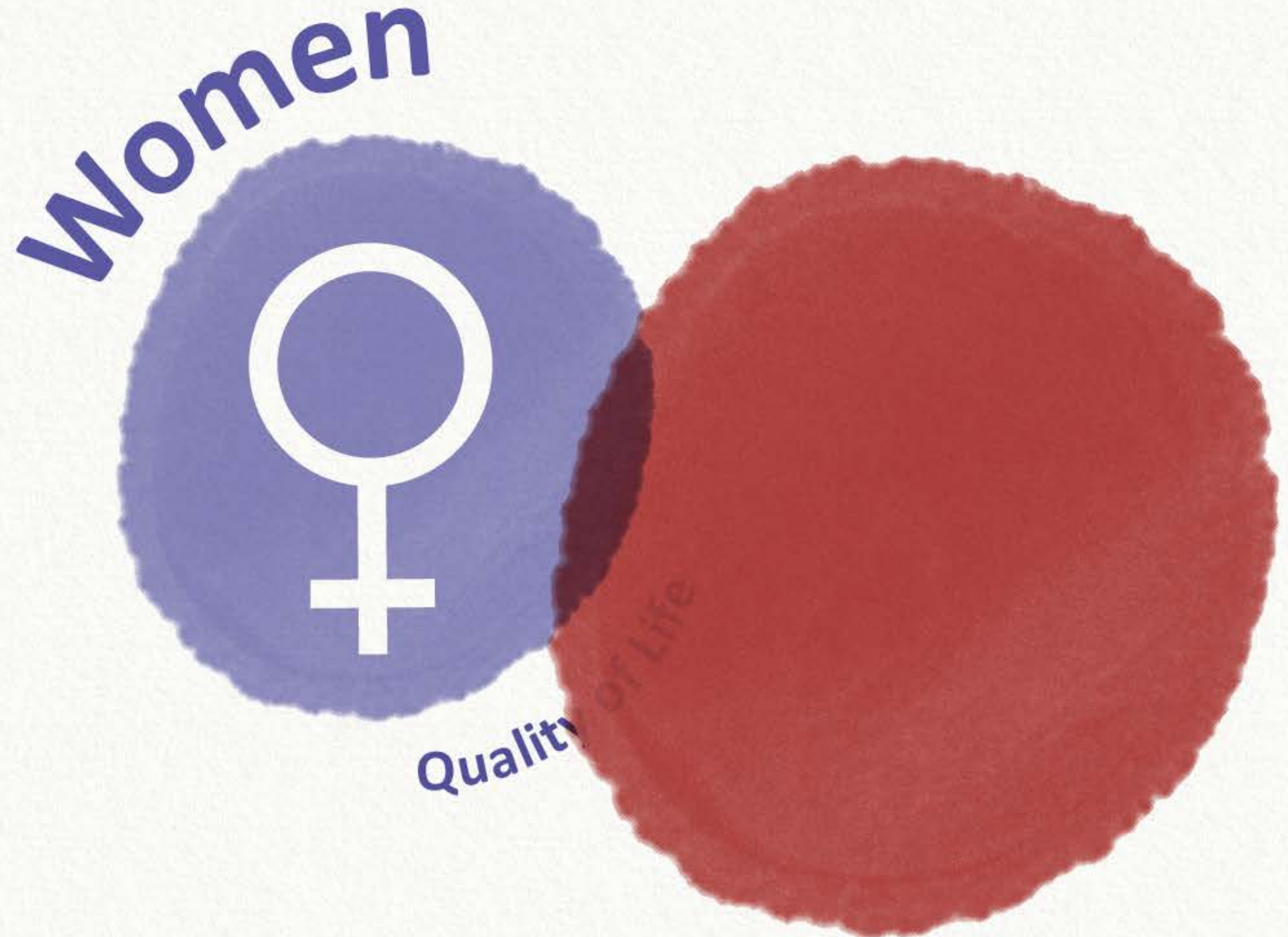
- Professor and Consultant Obstetrician and Gynaecologist at the Royal Free Hospital and University College London
- Leads multidisciplinary team service for women with bleeding and thrombotic disorders
- Leads research and postgraduate training on women and haemostasis
- Author of the book *Inherited Bleeding Disorders in Women* and coauthor of (inter)national guidelines and more than 200 publications

Bleeding disorders do occur in women and girls and have a **major impact on quality of life**

Bleeding disorders occur **as often in women as in men**, but women experience the additional bleeding challenges of menstruation and childbirth.

These disorders have a **major impact on the quality of life and social participation** of women and girls.¹⁻³

- Studies show clinically and statistically significant reductions in both physical and mental aspects of health-related quality of life



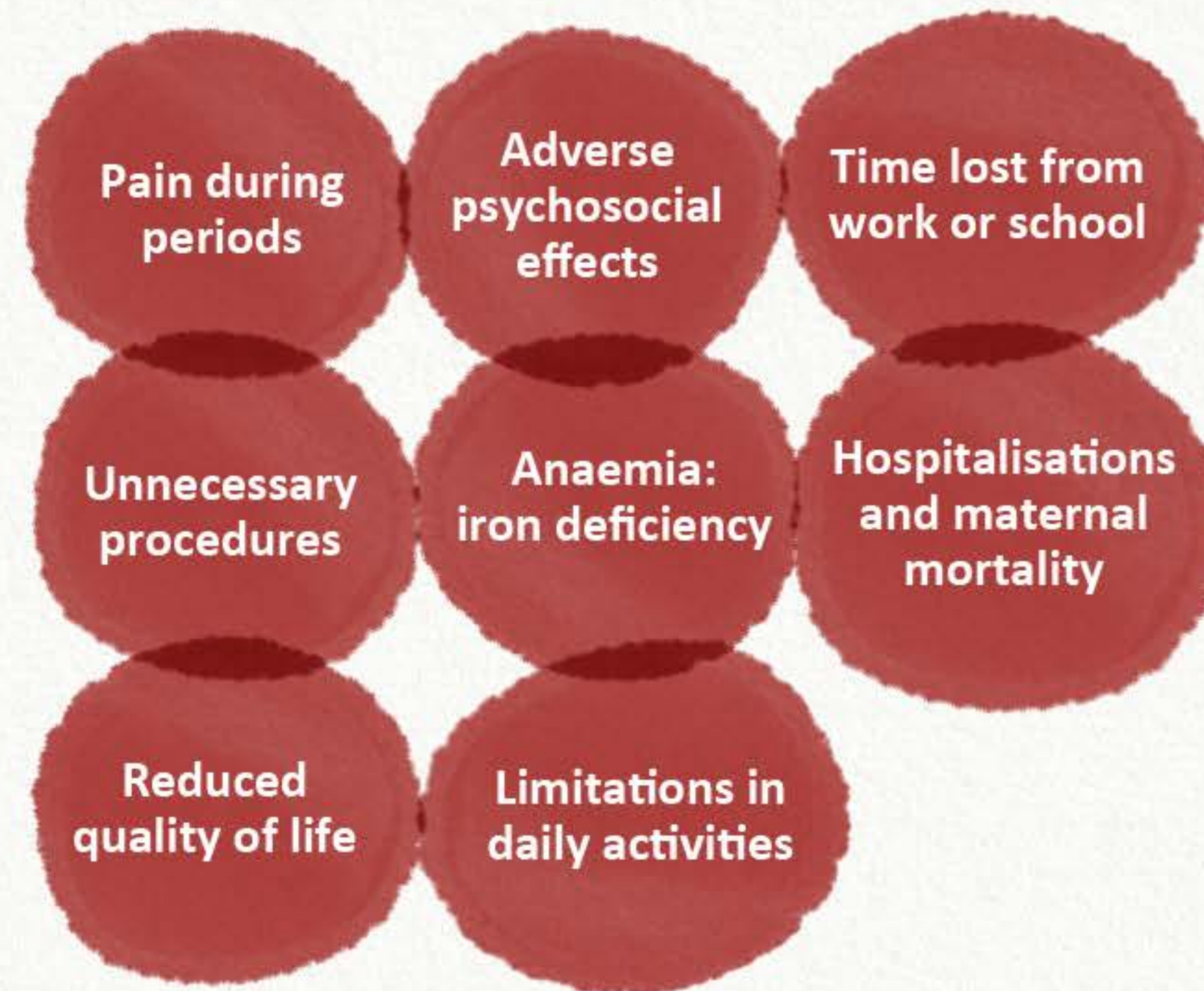
Bleeding disorders do occur in women and girls and have a **major impact on quality of life**

Heavy menstrual bleeding is a common symptom, and sometimes it is the only sign of a bleeding disorder.^{2,3}

- In adolescents, heavy menstrual bleeding is associated not only with iron deficiency (in up to 60% of patients) and fatigue but also with missed school days, social events, outdoor activities, and physical education⁴⁻⁶

Despite the negative impact of bleeding disorders on health-related quality of life, there is a long **delay** between the onset of symptoms and diagnosis of a bleeding disorder.^{3,7}

IMPACT OF BLEEDING DISORDERS ON WOMEN AND GIRLS¹



1. Kulkarni R. Eur J Haematol. 2015;95 Suppl 81:2-10; 2. Lavin M, et al. Blood Adv. 2018;2:1784-91; 3. Ragni MV, et al. Haemophilia. 1999;5:313-7; 4. Srivaths L, et al. Blood Adv. 2020;4:3209-16; 5. Wang W, et al. Haemophilia. 2013;19:225-30; 6. Torres R, et al. Rev Chil Pediatr. 2017;88:717-22; 7. Noone D, et al. Haemophilia 2019;25:468-74

Bleeding symptoms can be caused by a **variety of disorders** in **platelets and blood clotting factors**

HAEMOSTASIS CONSISTS OF THREE PHASES

1. VASOCONSTRICTION



Vasoconstriction is generally not affected by bleeding disorders

2. PLUG FORMATION BY PLATELETS



Bleeding disorders in women

- Von Willebrand's disease
- Congenital abnormalities of platelet count and function

3. CLOT FORMATION IN THE COAGULATION CASCADE



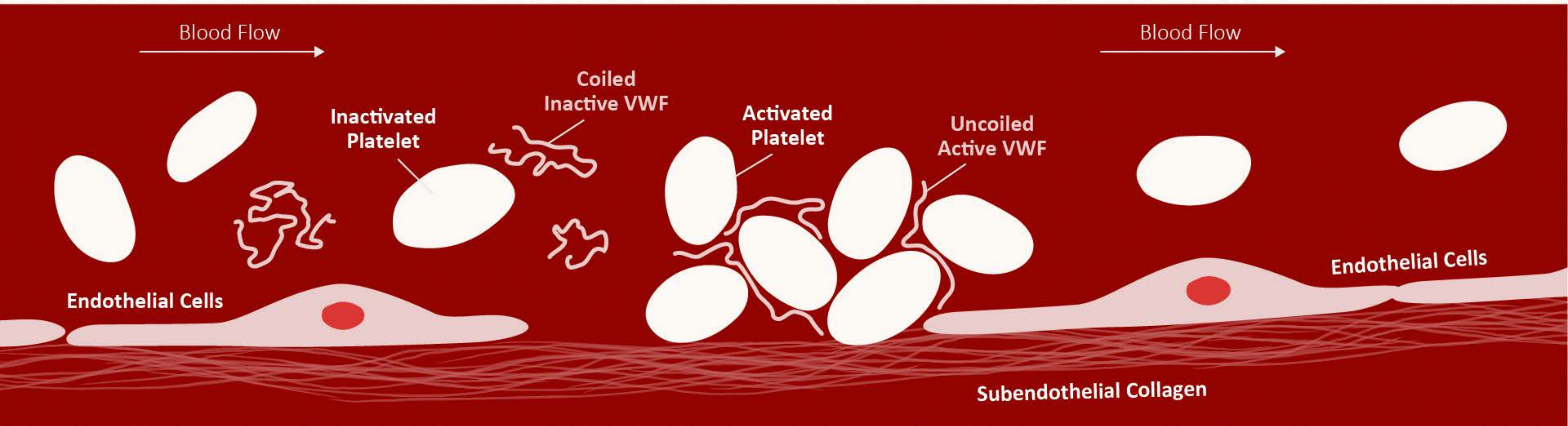
Bleeding disorders in women

- Haemophilia carriership
- *Rare bleeding disorders*

Plug formation can be affected by bleeding disorders in women and girls

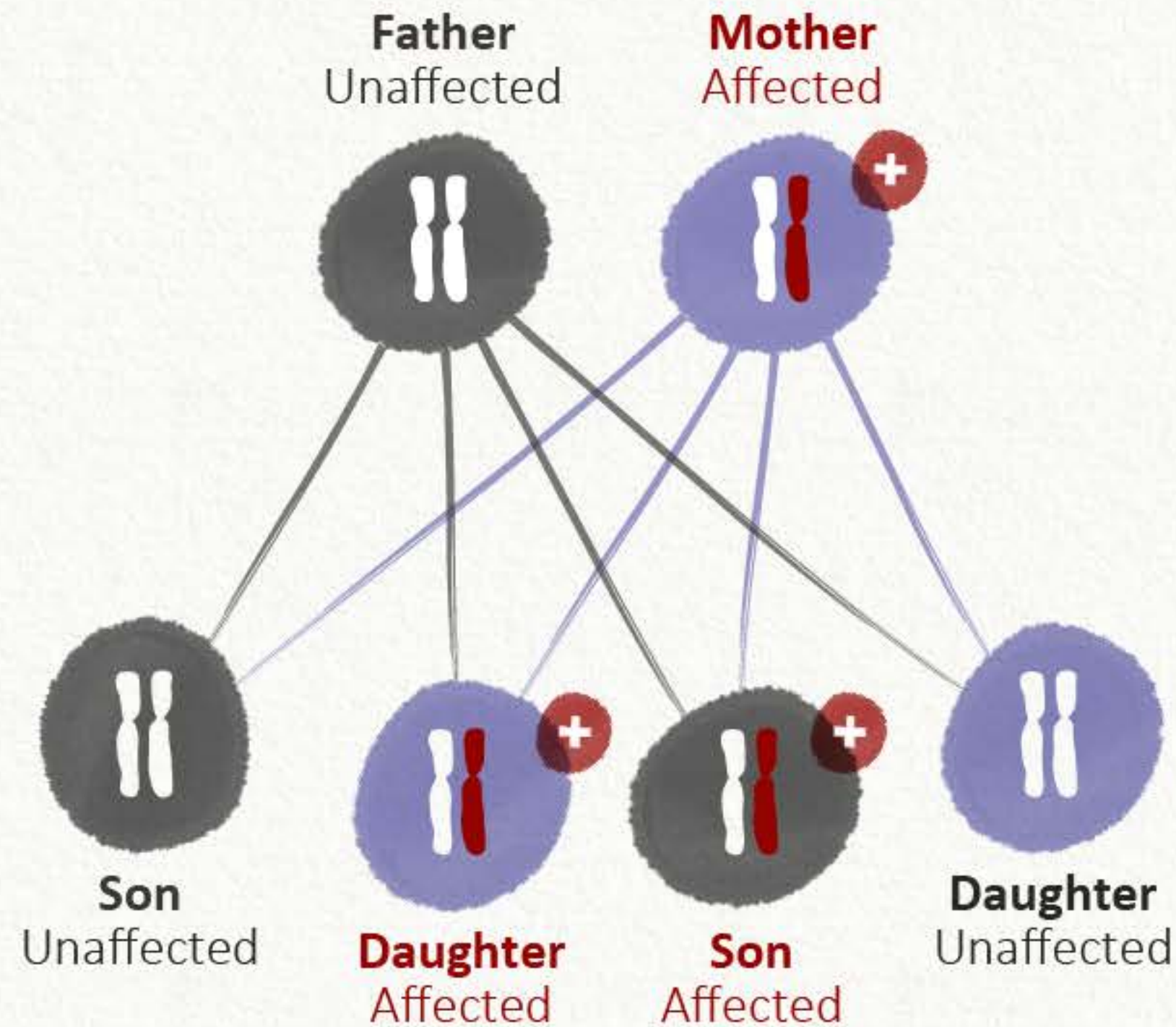
Von Willebrand factor (VWF) binds to collagen at sites of vascular injury and mediates platelet adhesion and aggregation. Patients with **Von Willebrand's disease** have reduced levels of (functional) VWF and, therefore, problems in plug formation. Von Willebrand's disease is the **most common inherited bleeding disorder**.

Congenital abnormalities of platelet count and function are a heterogeneous group of disorders that can result in bleeding symptoms ranging from mild bruising to severe haemorrhage.



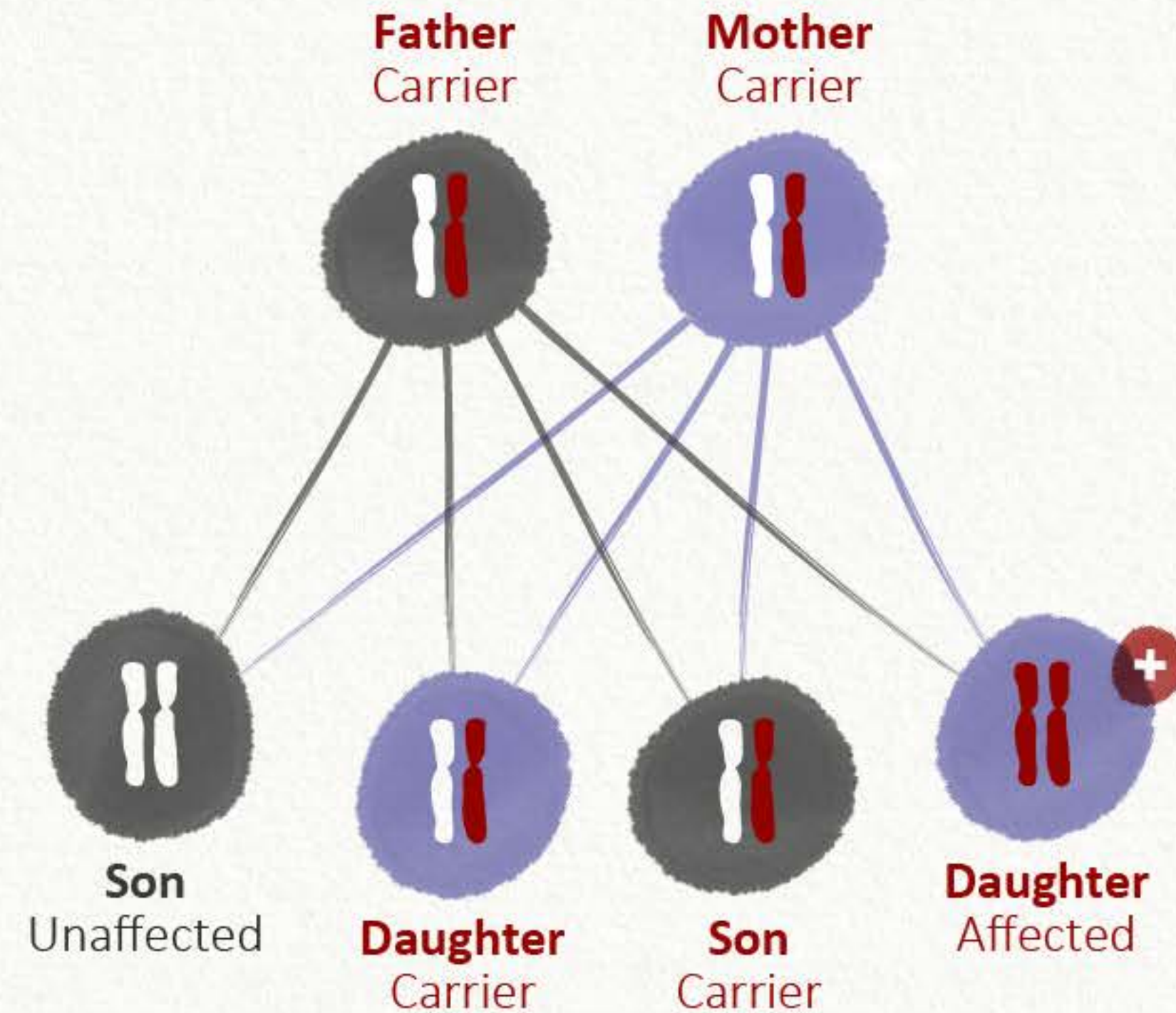
Plug formation can be affected by bleeding disorders in women and girls

AUTOSOMAL DOMINANT



Von Willebrand's disease and congenital platelet defects typically have an **autosomal dominant** pattern of inheritance.

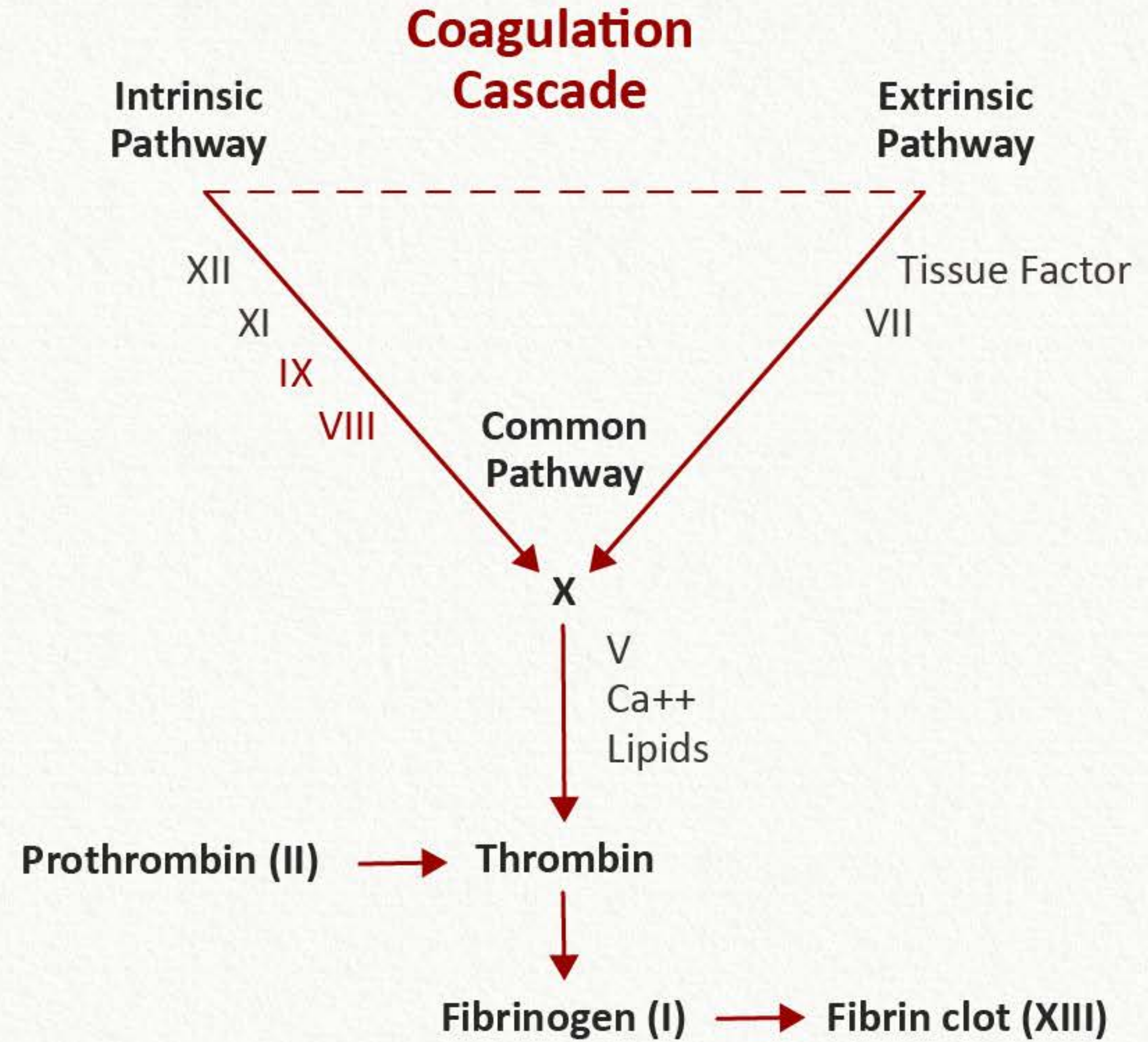
AUTOSOMAL RECESSIVE



Type 3 Von Willebrand's disease and most severe types of platelet function defects have an **autosomal recessive** pattern of inheritance.

The **coagulation cascade** can be affected by bleeding disorders in women and girls

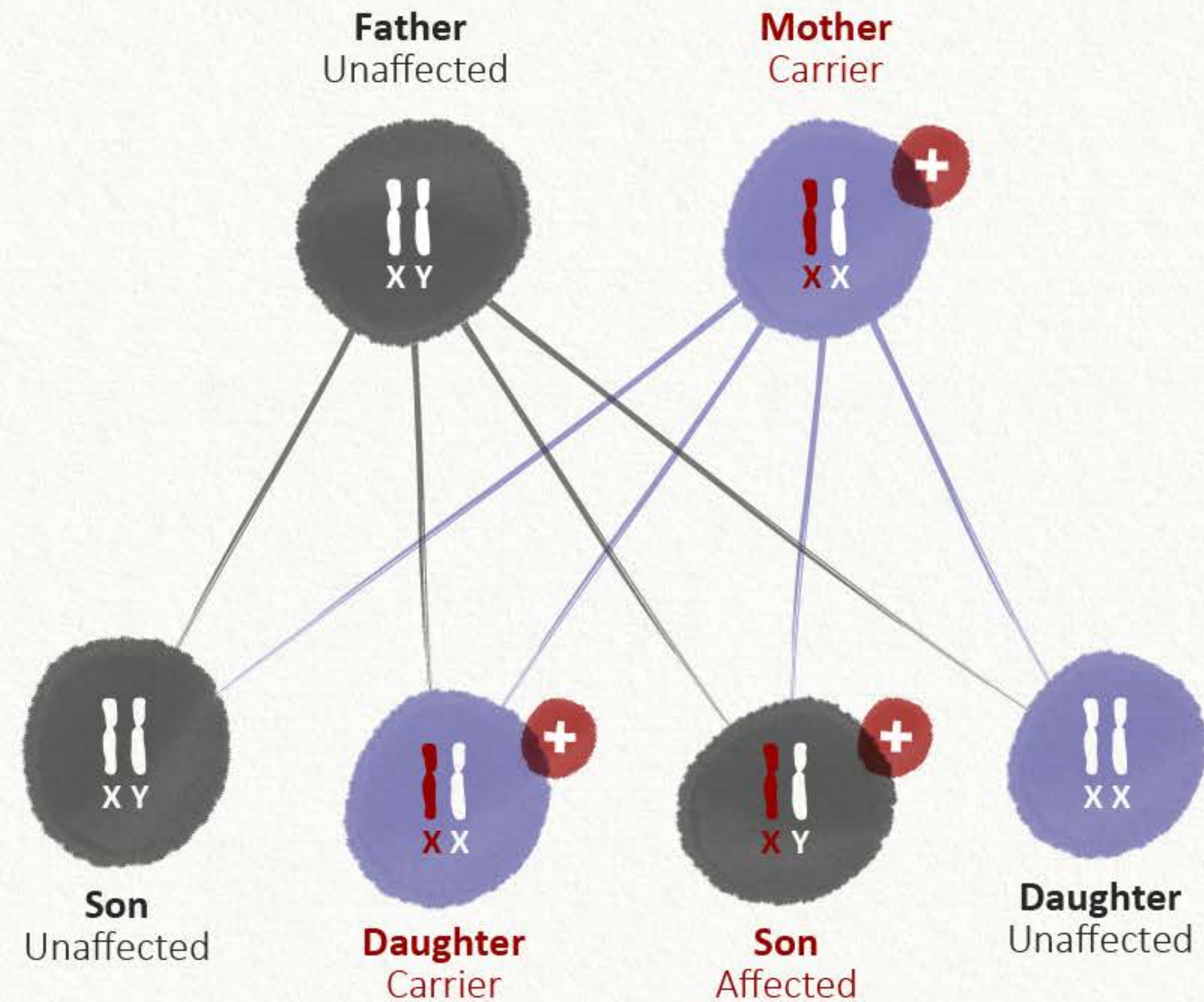
Haemophilia is a hereditary bleeding disorder characterised by deficiency of coagulation factor VIII (haemophilia A) or factor IX (haemophilia B).¹ Haemophilia A is more common than haemophilia B.



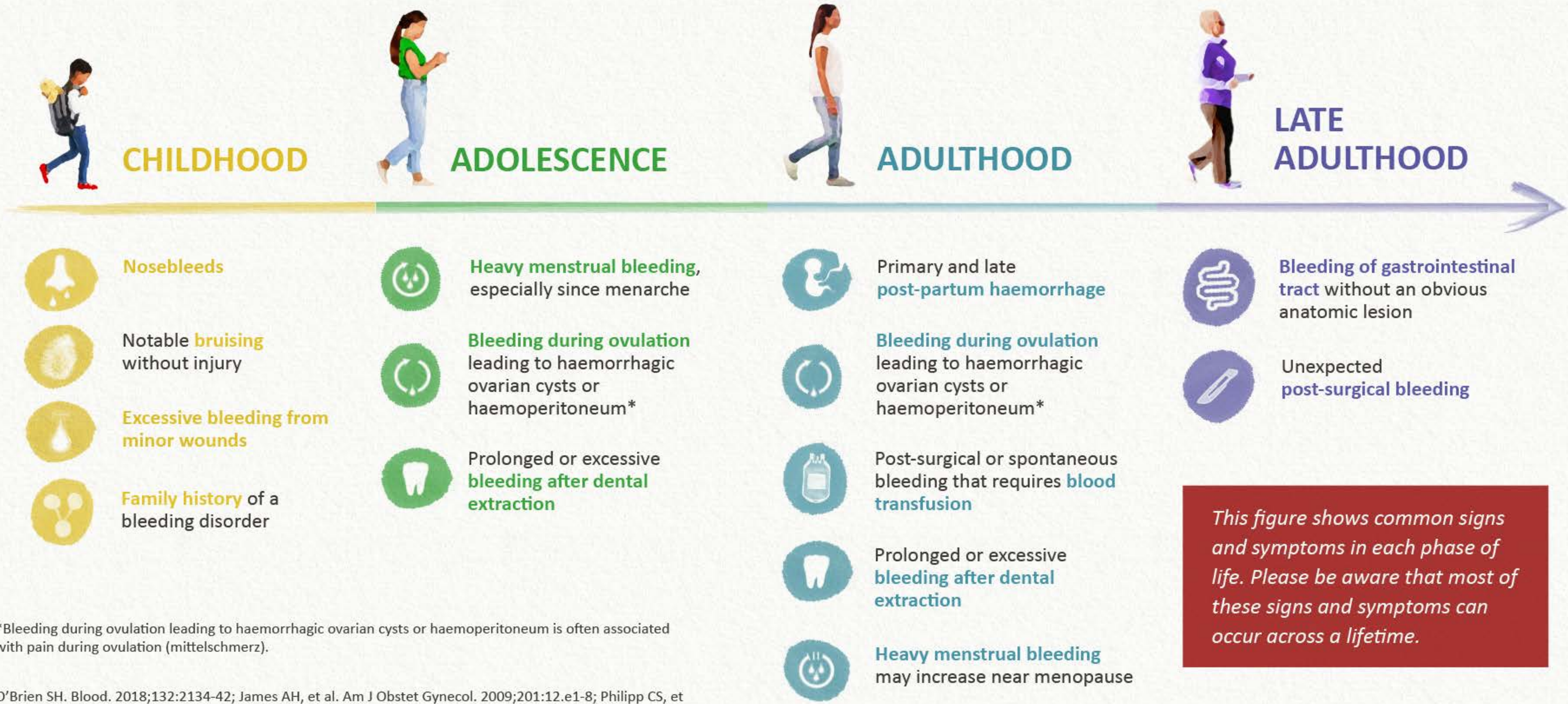
The **coagulation cascade** can be affected by bleeding disorders in women and girls

Haemophilia has an **X-linked recessive** pattern of inheritance.¹ Women and girls with an altered copy of the gene on chromosome X are called **carriers**. Approximately 1/3 of haemophilia carriers exhibit **low factor levels**.

! Carriers often have a clinically relevant increased bleeding tendency, even if their factor levels are normal.^{2,3}



Signs and symptoms suggesting an underlying bleeding disorder in women and girls



*Bleeding during ovulation leading to haemorrhagic ovarian cysts or haemoperitoneum is often associated with pain during ovulation (mittelschmerz).

O'Brien SH. Blood. 2018;132:2134-42; James AH, et al. Am J Obstet Gynecol. 2009;201:12.e1-8; Philipp CS, et al. Obstet Gynecol. 2005;105:61-6

This figure shows common signs and symptoms in each phase of life. Please be aware that most of these signs and symptoms can occur across a lifetime.

Identifying women and girls who are likely to have a bleeding disorder

Front-line healthcare professionals play a crucial role in **reducing the time to diagnosis** of bleeding disorders in women and girls.

Assessing the **personal bleeding history and family history** is key to identifying women and girls who are likely to have undiagnosed bleeding disorders.

One of the following **screening tools** can be used to screen for a bleeding disorder by:

- Pictorial blood assessment chart
- Philipp tool
- ISTH-SSC bleeding assessment tool

PARTICULARLY CONSIDER SCREENING FOR BLEEDING DISORDERS IN WOMEN AND GIRLS WITH



Heavy menstrual bleeding, especially since menarche



Iron deficiency or iron-deficiency anaemia due to gynaecological bleeding



Family history of bleeding symptoms



Primary or late post-partum haemorrhage



Recurrent ovulation bleeding



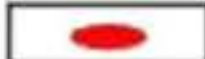





Multiple bleeding symptoms

The **pictorial blood assessment chart** allows patients to quantify blood loss during menstruation

DETECTING HEAVY MENSTRUAL BLEEDING

Patients fill out the chart themselves during their period by placing a tally mark, each time they change their sanitary protection, next to the amount of bleeding as well as recording clots and any cases of flooding.

At the end of their period, patients calculate the total score by multiplying the number of tallies in each row by the multiplying factor and adding up to get the row total.

DAY	DAY1	DAY2	DAY3	DAY4	DAY5	DAY6	DAY7	DAY8	DAY9	DAY10	TOTAL TALLIES	MULTIPLYING FACTOR	ROW TOTAL
												X1	
												X5	
												X20	
												X1	
												X5	
												X10	
Small blood clots (= Dime)												X1	
Large blood clots (≥ Quarter)												X5	
Menstrual accidents												X5	
Total Score (Sum of rows)													

A total score (sum of row scores) of ≥100 indicates heavy menstrual bleeding

The **Philipp tool** identifies women and girls who need further evaluation

The Philipp tool was developed to help **identify women for testing and further evaluation** for bleeding disorders.

The Philipp tool is considered **positive** if a response to 1 or more of the questions in the severity category reveals abnormality and/or an affirmative response is obtained in any other category.

Philipp tool

SEVERITY OF MENORRHAGIA

- Q1. How many days did your period usually last, from the time bleeding began until it completely stopped?
Abnormal if ≥ 7 days
- Q2. How often did you experience a sensation of “flooding” or “gushing” during your period?
Abnormal if every or most periods
- Q3. During your period did you ever have bleeding where you would bleed through sanitary protection in 2 hours?
Abnormal if every or most periods

HISTORY OF TREATMENT FOR ANAEMIA

- Q4. Have you ever been treated for anaemia?

FAMILY HISTORY

- Q5. Has anyone in your family ever been diagnosed with a bleeding disorder?

PERSONAL HISTORY

- Q6. Have you ever had a tooth extracted or had dental surgery?
If yes, did you have problem with bleeding after tooth extraction or dental surgery?
- Q7. Have you ever had surgery other than dental surgery?
If yes, did you have bleeding problem after surgery?
- Q8. Have you ever been pregnant?
If yes, have you ever had bleeding problem after delivery or after a miscarriage?

ISTH-SSC bleeding assessment tool further assesses patients with a suspected inherited bleeding disorder

The ISTH-SSC subcommittees have developed a tool to **assess patients with a suspected inherited bleeding disorder** — it can be used for men and for women.

The questionnaire is to be **collected by an adequately trained healthcare professional**, who will score symptoms and related treatments before or at diagnosis (or both).

In women, a **score of ≥ 6 is considered abnormal** and indicates that further testing is required.

The ISTH-SSC bleeding assessment tool scores the following symptoms

- | | |
|-------------------------------|-------------------------------------|
| 1. Epistaxis | 9. Menorrhagia |
| 2. Cutaneous bruising | 10. Post-partum haemorrhage |
| 3. Bleeding from minor wounds | 11. Muscle haematomas |
| 4. Oral cavity bleeding | 12. Haemarthrosis |
| 5. Gastrointestinal bleeding | 13. Central-nervous system bleeding |
| 6. Haematuria | 14. Other bleeding problems |
| 7. Dental extraction bleeds | |
| 8. Surgical bleeding | |



An online version of the ISTH bleeding assessment tool is available at <https://bleedingscore.certe.nl>

Next steps when suspecting a bleeding disorder



1

Start initial treatment

Hormonal therapy or tranexamic acid (or both) for heavy menstrual bleeding

Iron replacement therapy for iron deficiency

2

Consult a haematologist

for diagnostic laboratory assessment

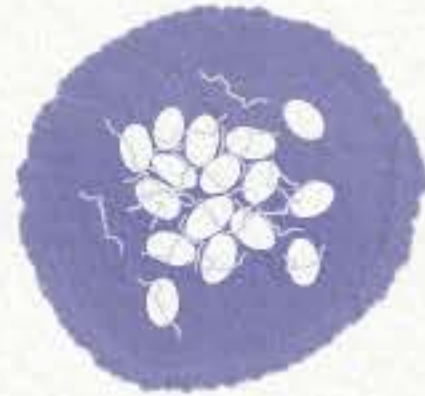


For more information on the diagnosis and management of bleeding disorders, please refer to Module 2 of this micro e-learning programme at www.checkpoint.cor2ed.com

Summary



Bleeding disorders occur as often in women as they do in men and have a major impact on quality of life



Bleeding symptoms can be **caused by a variety of disorders** in platelets and blood clotting factors



Signs and symptoms suggesting a bleeding disorder in women and girls include long-standing heavy menstrual bleeding, post-partum bleeding, and iron-deficiency anaemia



Assessing the **personal bleeding history and family history** are key to identifying women and girls who are likely to have undiagnosed bleeding disorders and should undergo laboratory evaluation



When suspecting a bleeding disorder, **start initial treatment and refer to a haematologist** for diagnostic laboratory assessment

Next steps

Please now proceed to the **assessment quiz** to test your knowledge.

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

- The strengths and limitations of tests to diagnose bleeding disorders in women
- Basic interpretation of test results to diagnose bleeding disorders in women
- The management of bleeding symptoms in women until specialist assessment is available

Note: you will be able to claim your CME credit after passing the assessment quizzes in both Modules 1 and 2



Visit the assessment quiz at www.checkpoint.cor2ed.com

*CME credit available
Visit programme home page on:*

