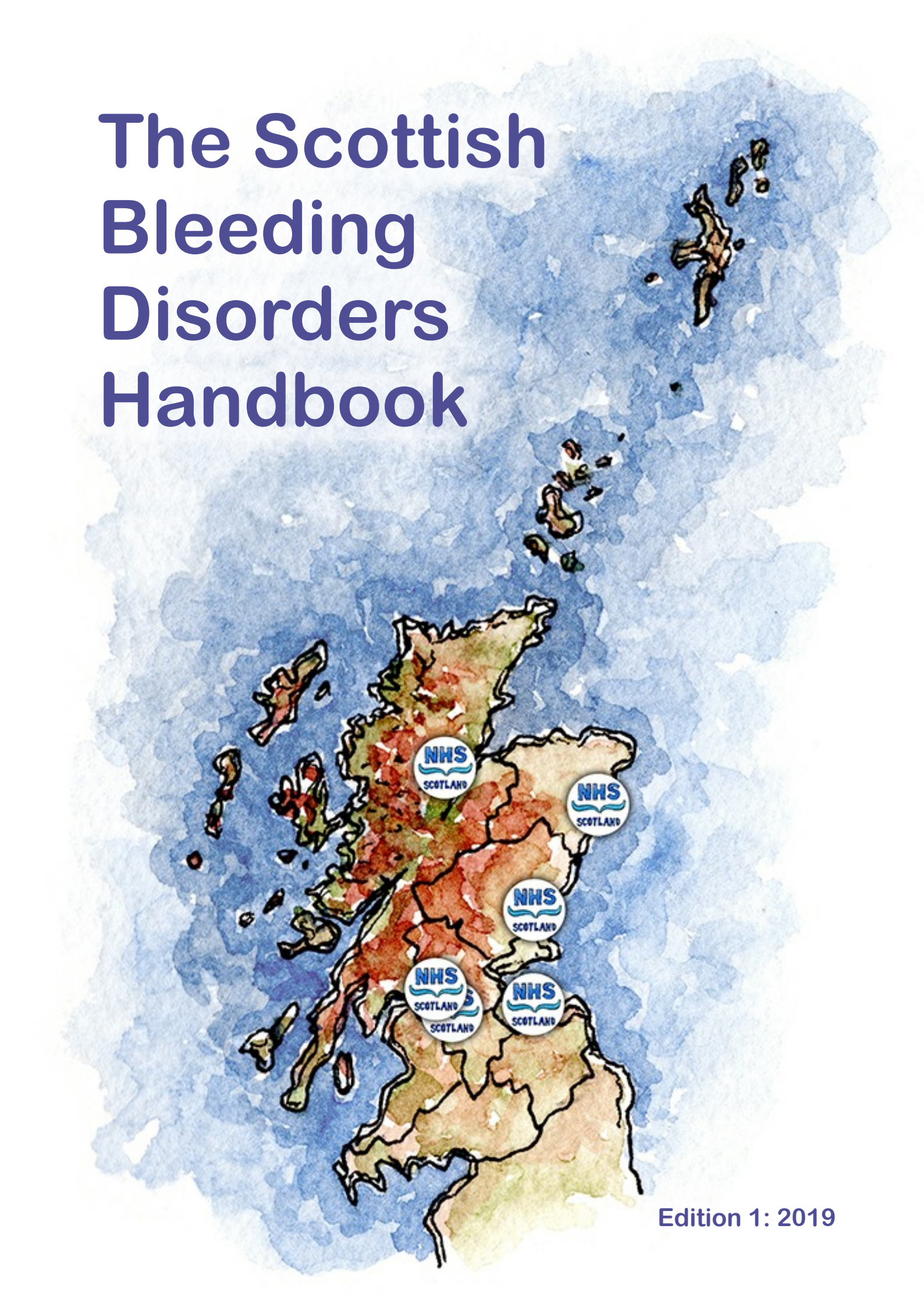
The Scottish Bleeding Disorders Handbook



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Acknowledgement

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Disclaimer

None of the information in this booklet is intended to replace medical advice. Haemophilia Scotland cannot be held responsible for any inaccuracies. Personal stories are representative examples only.

Other sources of information and support

Bleeding disorders

Scottish Inherited Bleeding Disorders Network www.mcns.scot.nhs.uk/sibdn
An extended multidisciplinary team including patients and carers that oversees the delivery of Scottish Haemophilia Services and coordinates all parties involved to achieve the best possible service
The Haemophilia Society UK www.haemophilia.org.uk
A UK charity with three local Haemophilia Groups in Scotland (Grampian, based in Aberdeen; Tayside, based in Dundee; West of Scotland, based in Glasgow)
World Federation of Hemophilia www.wfh.org
European Hemophilia Consortium www.ehc.eu
UK Haemophilia Centre Doctors' Organisation www.ukhcdo.org

Genetic/rare conditions

The Genetic Alliance www.geneticalliance.org.uk
Rare Diseases UK www.raredisease.org.uk

Charities and organisations working those affected by blood borne viruses

Scottish Infected Blood Support Scheme www.nhsnss.org/services/practitioner/medical/scottish-infected-blood-support-scheme
Provides support to people who were historically infected with hepatitis C and/or HIV following treatment with NHS blood, blood products or tissue.
The Hepatitis C Trust (Scotland) www.hepctrust.org.uk/scotland
The Scottish Infected Blood Forum www.sibf.org.uk
Hepatitis Scotland www.hepatitisscotland.org.uk
HIV Scotland www.hivscotland.com
Waverley Care (for HIV and hepatitis C) www.waverleycare.org

Introduction

Who is this booklet for?

Haemophilia Scotland has produced this handbook to help all people in Scotland who are affected by an inherited bleeding disorder. You could be newly diagnosed or have known about your condition for a long time and want to explain it to others. Alternatively you might be working at a school or nursery, or be a childminder or run a sports club, and be responsible for someone with a bleeding disorder.

If you need to find out more about haemophilia, von Willebrands or any of the other bleeding disorders, then this handbook is for you!

Contents

Haemophilia Scotland

Haemophilia Scottland is a Scottish registered charity (no. SC044298) for people who have haemophilia, von Willebrands and other inherited bleeding disorders.

Through talking to all sections of the patient community of individuals and families with inherited bleeding disorders, we form a united representative voice when speaking with the healthcare professionals, government and the rest of Scottish society, and when connecting with people like us throughout the world.

The Scottish Inherited Bleeding Disorders Network

The purpose of the Scottish Inherited Bleeding Disorder Network (SIBDN) is to facilitate clinical and other improvements for individuals with inherited bleeding disorders.

A key aim of the Network is to enable timely and effective care for individuals with inherited bleeding disorders across Scotland.

Ten things you need to know about inherited bleeding disorders

1. **Inherited bleeding disorders** are lifelong conditions where your blood doesn't clot properly, causing you to bleed longer.

2. All the bleeding disorders described in this handbook are caused by genes that are passed from parents to children (i.e. **inherited**). Sometimes they are diagnosed in an infant when there's no known family history of bleeding.

3. **Haemophilia** is the most well-known type of bleeding disorder. Others include: **von Willebrands**, **clotting factor deficiencies** and **platelet disorders**.

4. The medical study of bleeding disorders is a branch of **haematology and haemostasis**. They are managed in NHS **Haemophilia Comprehensive Care Centres** or smaller **Haemophilia Centres**.

5. There are now good treatments for most bleeding disorders to help stop bleeding (**on demand treatment**). Regular continuous treatment (**prophylaxis**) can also protect you from future bleeds.

6. Your pattern of bleeding depends on the type of bleeding disorder. **Surface (superficial) bleeds** include cuts that bleed a lot after an accident, dental work or surgery, and gum and nose bleeds. **Internal bleeds** into the **brain** or **stomach** are very serious and can be life-threatening, whilst bleeds into **joints** and **muscles** are very painful and can cause swelling, bruising and long-term joint damage.

7. **Females** are just as likely to have a bleeding disorder gene as males, and face the extra challenges of heavy periods and bleeding after giving birth.

8. The word **P.R.I.C.E.** describes immediate first aid for bleeds into a joint or muscle (usually an elbow, knee or ankle): **P**rotection, **R**est, **I**ce, **C**ompression, **E**levation. Use treatment as quickly as possible.

9. Certain painkillers, particularly **aspirin** and **ibuprofen**, and a few other drugs, should be avoided.

10. With the treatment and care available in Scotland you can expect to lead a **normal life** and live as long as anyone without a bleeding disorder.

Myth Busting

FALSE If you cut yourself, even if it's small, you will lose a lot of blood.

TRUE You may bleed longer, but not any faster. Normal first aid measures should stop the bleeding.

FALSE Women can't have any symptoms of haemophilia.

TRUE Around a third of women who are carriers of haemophilia can have symptoms of mild or even moderate bleeding.

FALSE Only women pass the haemophilia gene onto their children.

TRUE All the daughters of a man with haemophilia will be carriers, but none of his sons will be affected.

FALSE Children with a severe form of bleeding disorder need to go to a special school.

TRUE Whilst some special measures may be needed, we recommend you should be able to choose any school.

FALSE You should avoid all physical exercise if you have a severe bleeding disorder.

TRUE Physical exercise helps build strength in joints and prevent painful bleeds into those joints. However, during a bleed you should rest the affected area.

FALSE Treatments for haemophilia and other bleeding disorders are unsafe.

FALSE You can catch von Willebrands.

TRUE In the past some people were infected with HIV and viral hepatitis from contaminated treatment, but the use of genetically-engineered (recombinant) products (where available) has eliminated this risk.

TRUE You can't 'catch' any of the bleeding disorders covered in this handbook (even those with 'disease' in their name). They are all inherited from a parent.

Introduction to bleeding disorders

There are many types of bleeding disorder described in this handbook. Haemophilia is the best known of them all, but von Willebrands is actually by far the most common. Some bleeding disorders only ever cause mild symptoms of bleeding, but others can be called moderate or severe.

The type of bleeding experienced depends on the bleeding disorder. For example, people with haemophilia often experience bleeding into joints and muscles, while people with von Willebrands tend to bleed more typically on the surface of their body (from nosebleeds and cuts to the skin and mucosal areas including gums).

How bleeding disorders are diagnosed

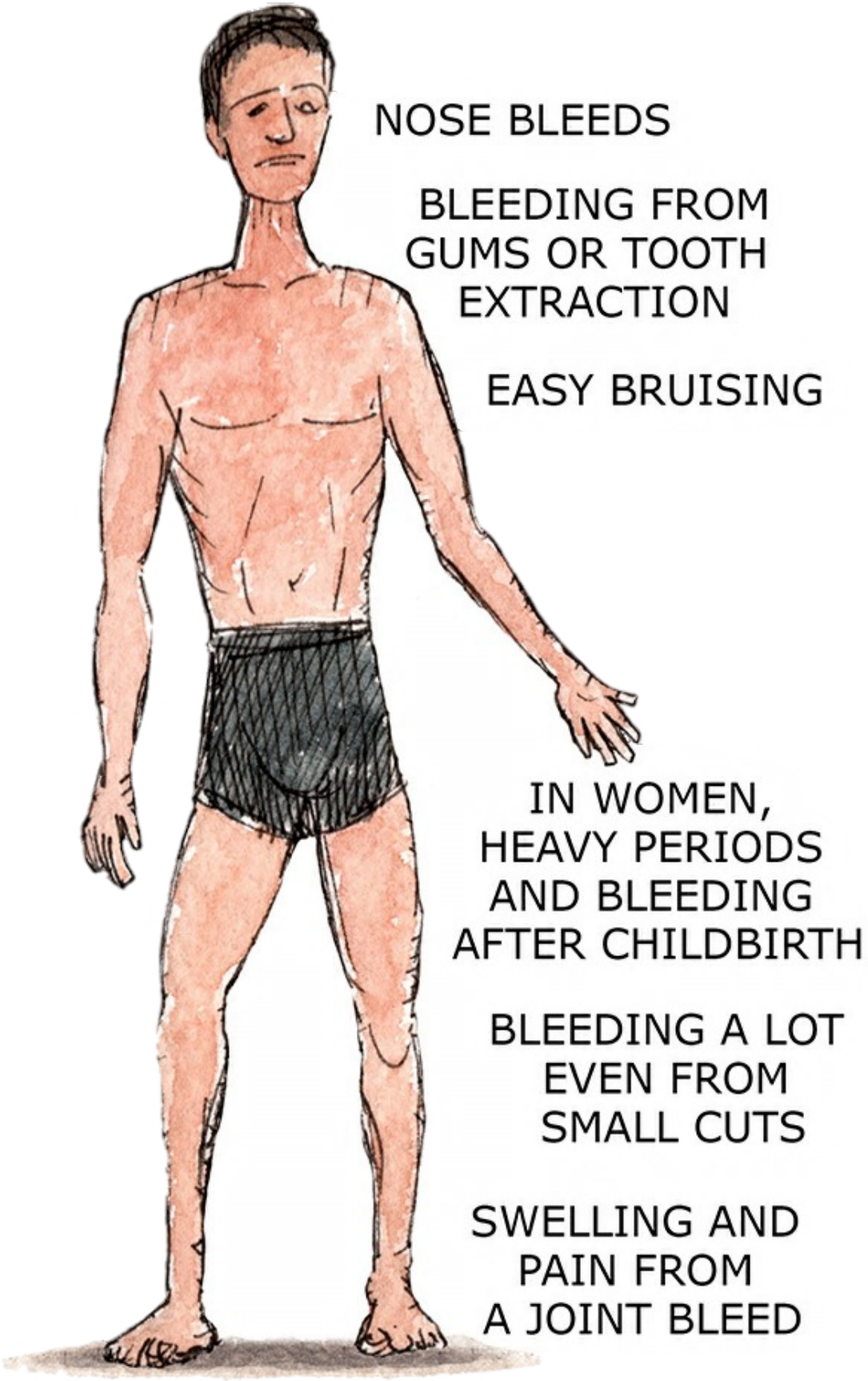
A combination of **family history** of bleeding, observing **symptoms** in the individual and **blood tests** enable your haemophilia specialist to diagnose your bleeding disorder.

If either biological parent has a bleeding disorder, their children may be affected. If there's an affected close family member, a bleeding disorder gene may be in the family.

Blood tests measure the ability of the blood to clot and detect any bleeding disorder genes.

In new-born babies, a bleeding disorder is suspected when there is excessive bleeding from the umbilical cord, after the heel-prick test, or after circumcision.

Common Symptoms of Bleeding Disorders

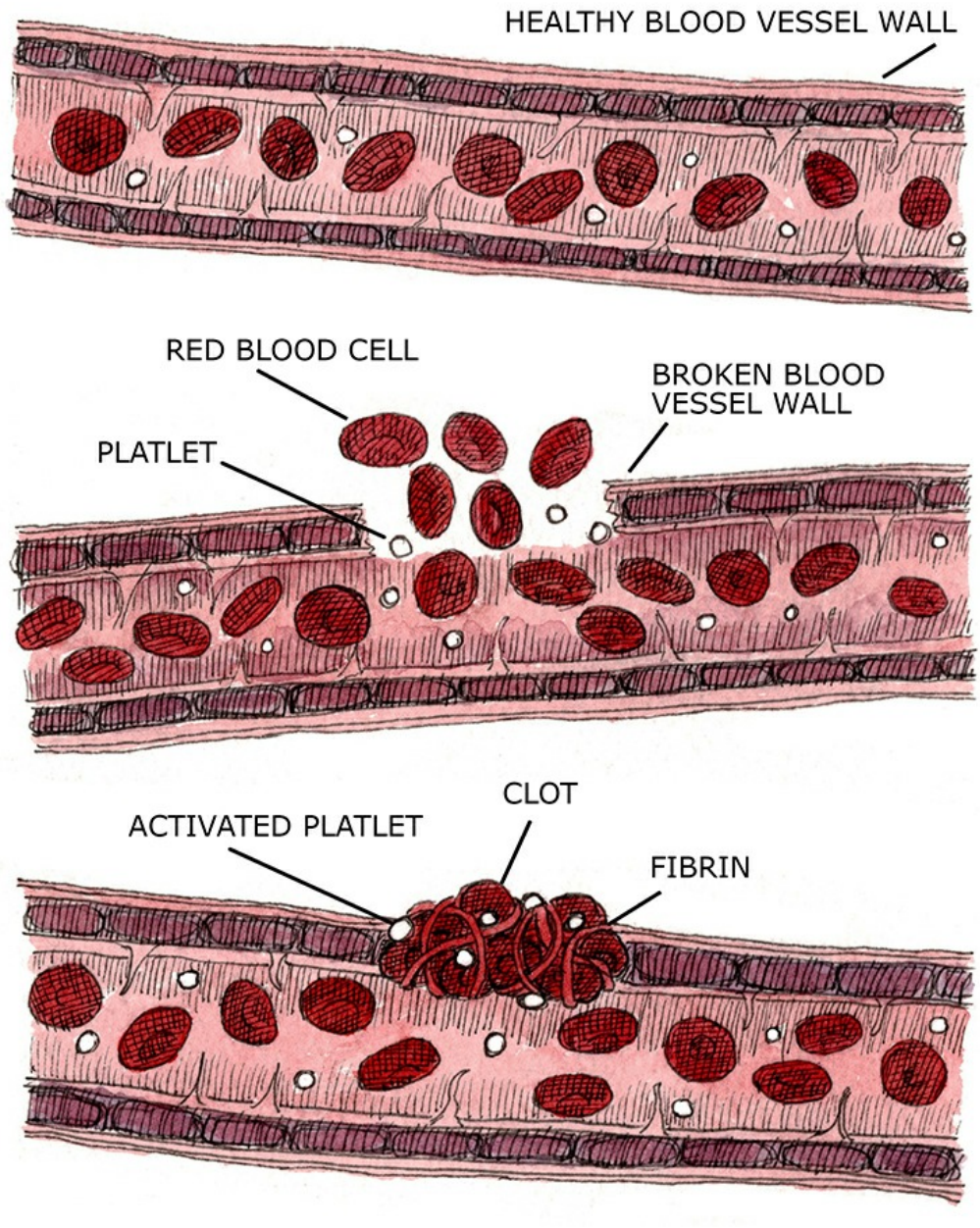


Bleeding and Clotting Explained

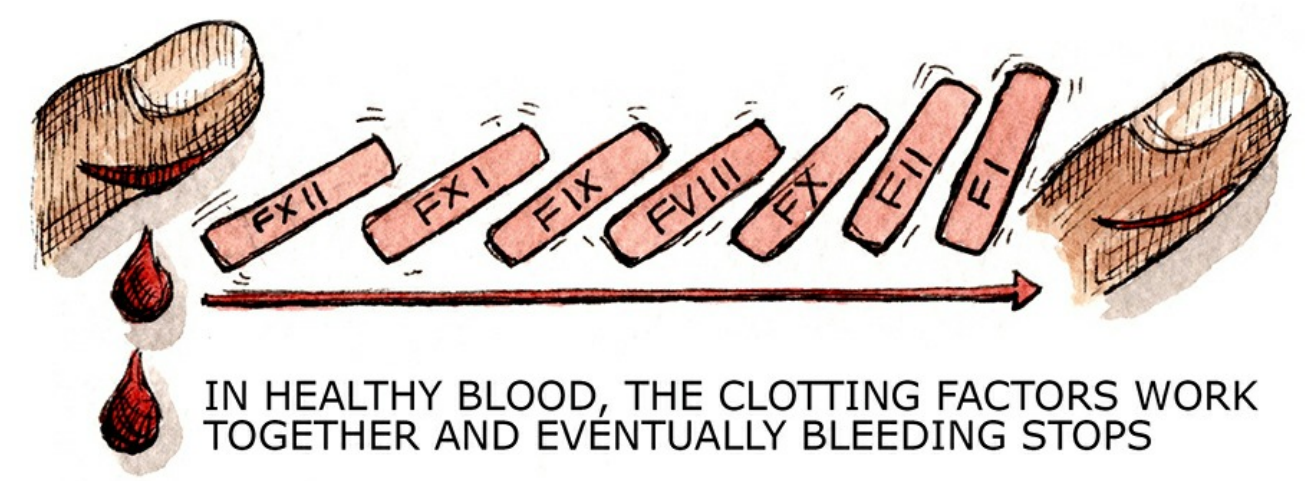
Blood has a number of functions of which the most important is supplying oxygen to all parts of your body via a vast network of blood vessels. Arteries take blood that is rich in oxygen away from the heart and lungs, and veins return it to the heart after the oxygen has been used.

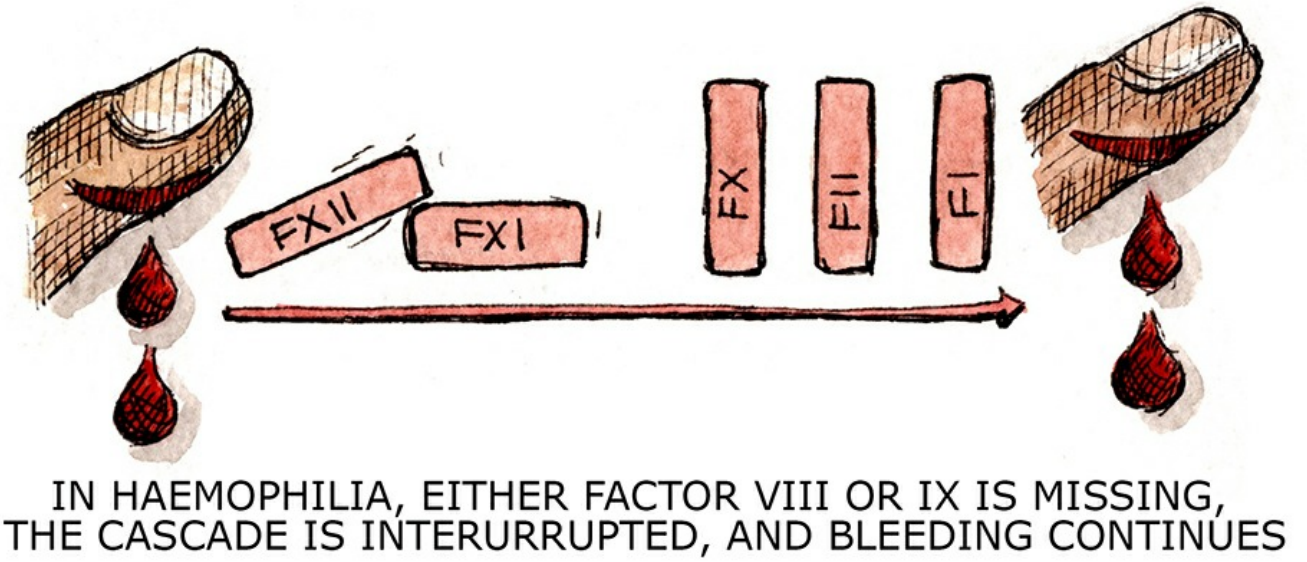
If a blood vessel is damaged and begins leaking, the body's 'puncture repair kit' is contained in both the blood vessel walls and the blood itself. A clot starts to form to repair the hole.

Platelets are small cells in the blood. Their main purpose is to help clotting, combining forces with a substance that leaks out the punctured blood vessel wall and proteins that are always present in the blood. Together, these form insoluble strands of a protein called fibrin, which binds together a clump of blood cells to form a clot that seals the hole and gives the blood vessel wall time to repair itself.



A **fibrin** clot is made at the end of a complex series of events involving twelve different blood clotting factors. This is much like a long line of dominoes where each one represents a clotting factor: if you knock the first one down each subsequent one falls in turn until they have all fallen, with the final one plugging a hole that represents a punctured blood vessel. If any clotting factor is low, absent or faulty, the domino cascade stops at that clotting factor, and the final hole is not plugged. In **haemophilia A** or **B** the 'weak' clotting factors are **Factor VIII (8)** or **IX (9)**, but a break in the cascade happens with all the other **clotting factor deficiencies**.





No bleeding disorder – the hole is plugged by the final domino and bleeding stops. Haemophilia or a clotting factor deficiency – the final domino fails to plug the hole and bleeding continues.

Haemophilia A and B

Haemophilia A or B occur when you have a shortage of a **clotting factor VIII (8) or IX (9)** in your blood, or when the clotting factor doesn't work properly. This means that whenever a blood vessel is damaged it is difficult for your blood to clot properly – it is slow to make the clot, and when a clot is formed it might not hold together very well.

If you have haemophilia A or B you tend to bleed more often than people without a bleeding disorder. The bleeding might be visible – such as to a cut or a nose or gum bleed – or may be internal, such as painful bleeds inside joints such as ankles and knees (which are the joints that bear your weight), or dangerous bleeds in the head or stomach.

Bleeds are usually treated by an injection of the relevant clotting factor concentrate – Factor VIII or IX. This is best given as soon as possible, and is known as **on demand** treatment. In addition, people with severe haemophilia A or B treat themselves every 1 to 14 days with clotting factor to help prevent bleeds occurring. This is known as **prophylaxis**.

It is not possible to tell the difference between haemophilia A and B without doing a blood test, but you need to know which type you have in order to use the right treatment.

The way your haemophilia is managed is very dependent on how severe it is. The amount of clotting factor VIII or IX in your blood will be measured and expressed as a percentage of the amount of an average person without haemophilia (they are said to have 50%-100% clotting factor).

Haemophilia A

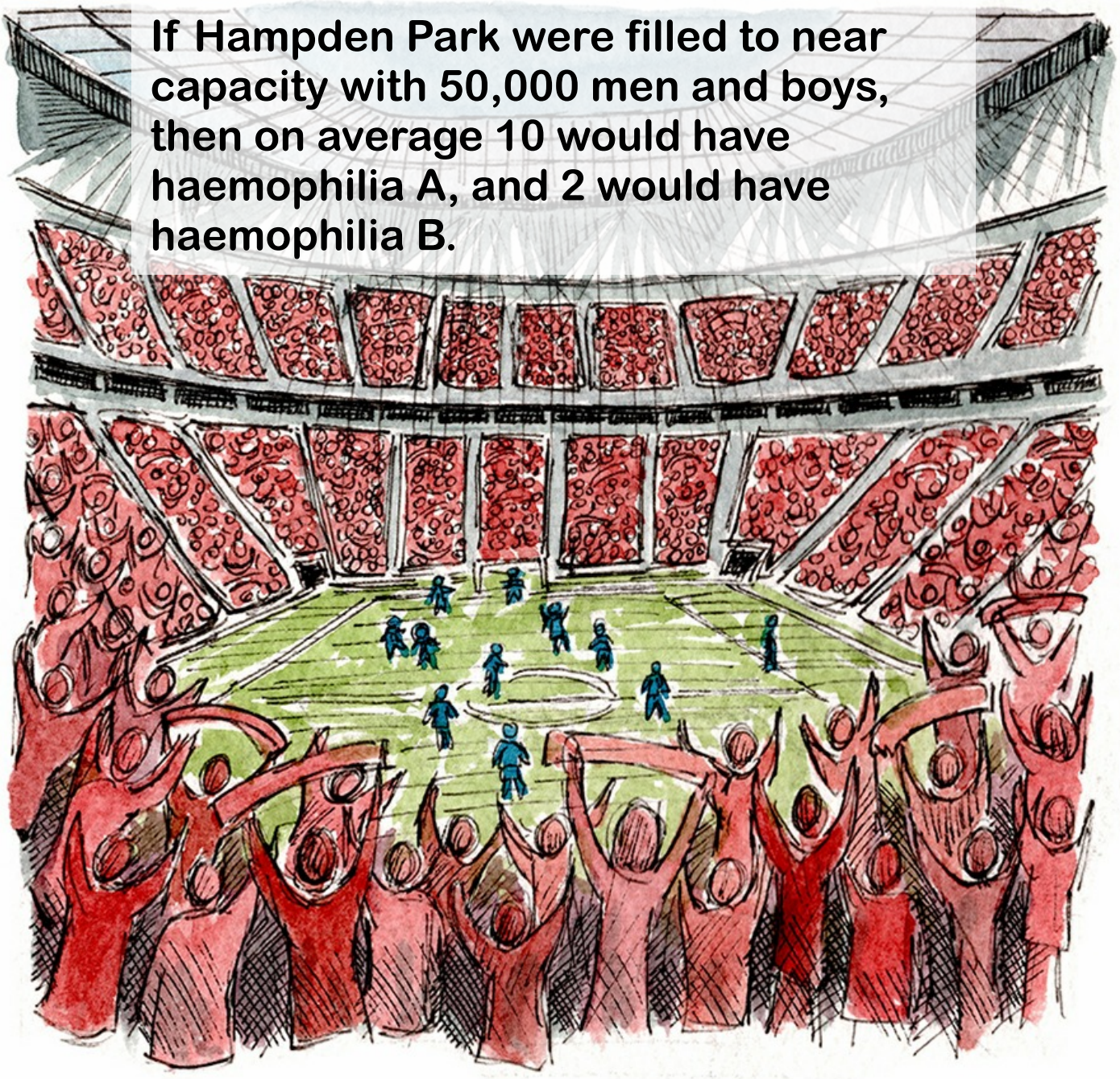
Severity range	Number of people in Scotland	Bleeding pattern	Inheritance	Other names
Severe (0 to less than 1% factor VIII)	147	Into joints, muscles, the head and after surgery	Sex-linked (see p 18)	• Congenital haemophilia • Factor VIII [8] deficiency
Moderate (1 to less than 5% factor VIII)	67			
Mild (5 to 40% factor VIII)	257			

Haemophilia B

Severity range	Number of people in Scotland	Bleeding pattern	Inheritance	Other names
Severe (0 to less than 1% factor IX)	24	Into joints, muscles, the head and after surgery	Sex-linked (see p 18)	• Christmas Disease • Factor IX [nine] deficiency
Moderate (1 to less than 5% factor IX)	43			
Mild (5 to 40% factor IX)	57			

Throughout the world, haemophilia A affects one in 5,000 males, and haemophilia B one in 25,000 males.

If Hampden Park were filled to near capacity with 50,000 men and boys, then on average 10 would have haemophilia A, and 2 would have haemophilia B.



Symptoms and management of haemophilia A and B		
	Symptoms	Management
Severe	Bleeding after injury and surgery, and joint bleeds for no apparent reason (spontaneous)	Prophylaxis and on demand treatment
Moderate	Bleeding after injury and surgery	On demand treatment. Prophylaxis normally only for surgery etc.
Mild	Bleeding after severe injury, and surgery	On demand treatment. Prophylaxis normally only for surgery etc.

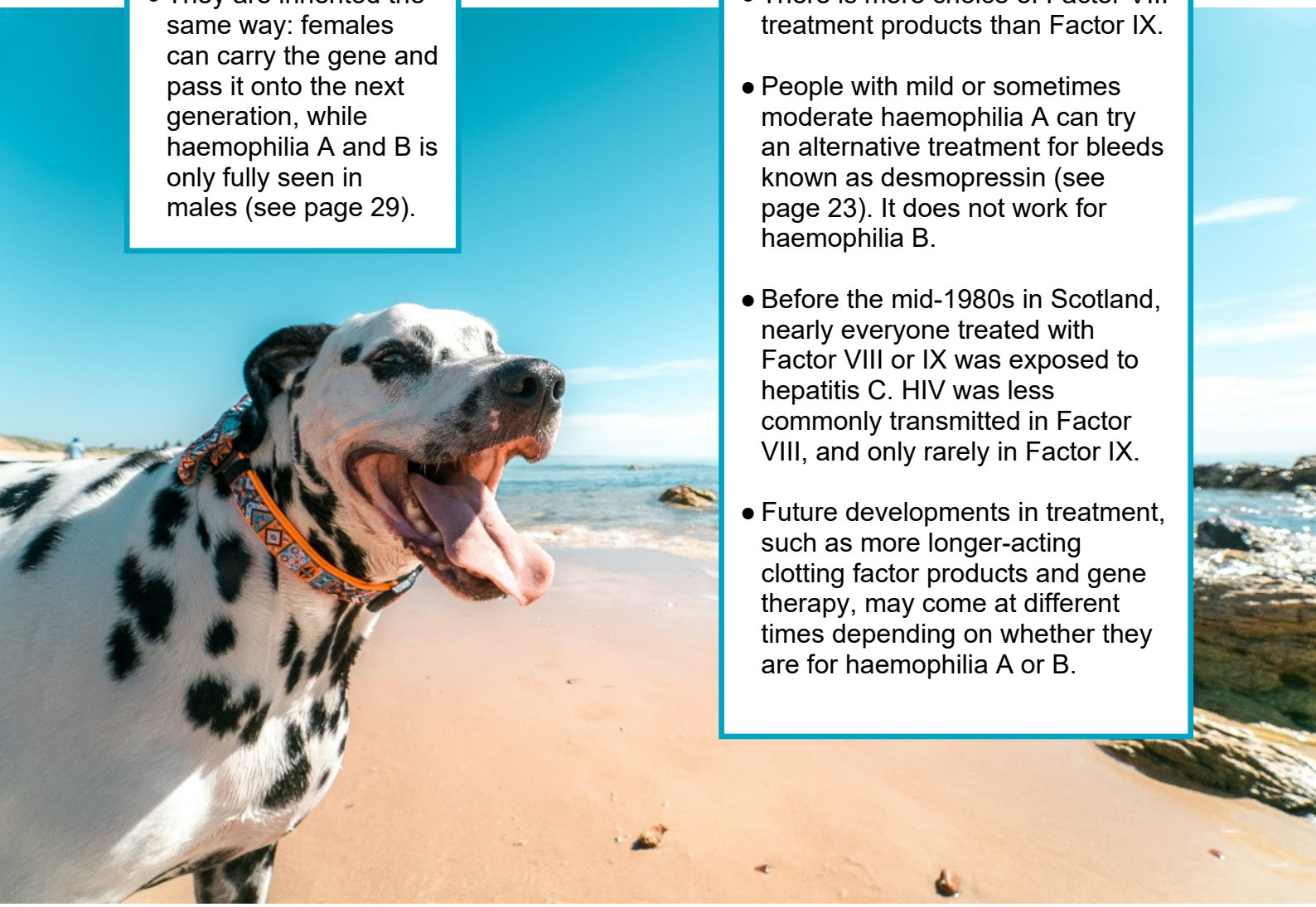
Spot the difference

Some similarities between haemophilia A and B

- Bleeding symptoms are the same, despite being caused by shortages of two different clotting factors.
- Readily-available treatments, now made by recombinant technology exist for both types.
- They are inherited the same way: females can carry the gene and pass it onto the next generation, while haemophilia A and B is only fully seen in males (see page 29).

Some differences between haemophilia A and B

- Prophylaxis in haemophilia B (factor IX) requires less infusions than in haemophilia A (factor VIII).
- If you have haemophilia B you are less likely to get an inhibitor (see page 21).
- Allergic reactions to Factor IX sometimes happen when first using it, but this is very rare with Factor VIII.
- There is more choice of Factor VIII treatment products than Factor IX.
- People with mild or sometimes moderate haemophilia A can try an alternative treatment for bleeds known as desmopressin (see page 23). It does not work for haemophilia B.
- Before the mid-1980s in Scotland, nearly everyone treated with Factor VIII or IX was exposed to hepatitis C. HIV was less commonly transmitted in Factor VIII, and only rarely in Factor IX.
- Future developments in treatment, such as more longer-acting clotting factor products and gene therapy, may come at different times depending on whether they are for haemophilia A or B.

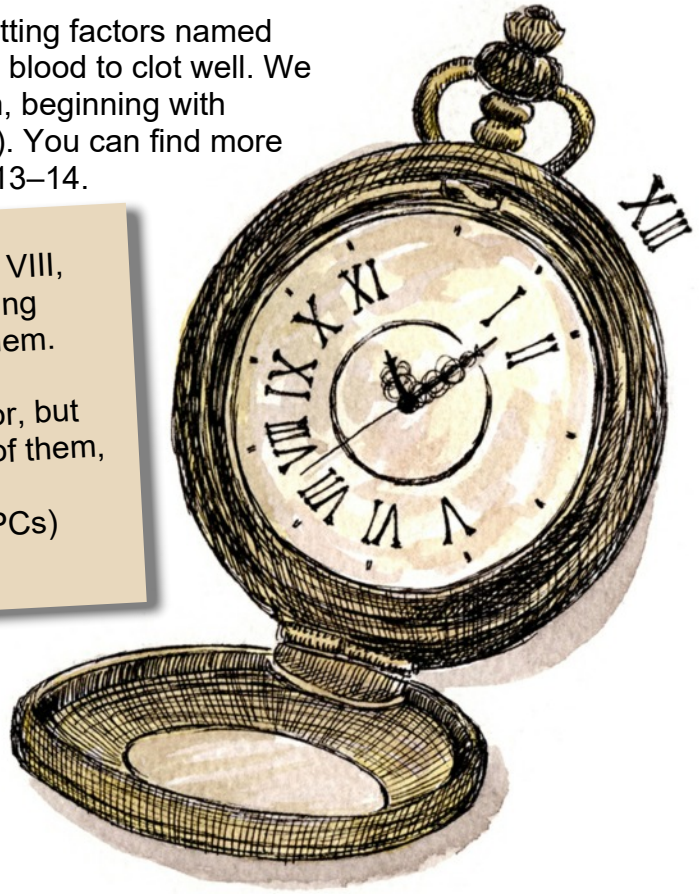


Other clotting factor deficiencies

On page 5 we saw how a set of a dozen clotting factors named with Roman numerals are necessary for the blood to clot well. We will look at each of these deficiencies in turn, beginning with fibrinogen (which can also be called factor I). You can find more information about the treatments on pages 13–14.

There are nine clotting factors (I, II, V, VII, VIII, IX, X, XI and XIII) that give rise to a bleeding disorder if you are lacking in any one of them.

Most treatments contain one clotting factor, but Fresh Frozen Plasma (FFP) contains all of them, while cryoprecipitate and the activated Prothrombin Complex Concentrates (aPPCs) contain some – see page 23.



The fibrinogen defects (also known as fibrinogen deficiency)

Type and range of severity	No. of people in Scotland	Inheritance	Bleeding pattern	Treatment
Afibrinogenaemia milder than haemophilia.	< 6	Both parents need to pass on gene	Bleeding after trauma, childbirth or surgery. Also thrombosis	Fibrinogen concentrate. Cryoprecipitate and tranexamic acid may also be used. Prophylaxis is possible.
Hypofibrinogenaemia milder than afibrinogenemia.	21	Either one or both of the parents need to pass on gene		Fibrinogen concentrate. Cryoprecipitate and tranexamic acid may also be used. Prophylaxis is possible.
Dysfibrinogenaemia 55% of people have no symptoms	199	Usually only one parent needs to pass on gene		Fibrinogen concentrate. Cryoprecipitate and tranexamic acid may also be used. Prophylaxis is possible.
Hypodysfibrinogenaemia severity of symptoms varies	12	Complex, but not sex-linked		No specific recommendations for treatment.

Prothrombin (Factor II) deficiency (hypoprothrombinaemia or dysprothrombinaemia)

Severity range	Number of people in Scotland	Bleeding pattern	Inheritance	Treatment options
Mild to severe. Carriers usually have no symptoms	< 6	Surface bleeds common, head bleeds, also joint and muscle bleeds.	Both parents need to pass on the gene	PCC (or FFP), tranexamic acid. Prophylaxis is possible.

*PCC contains Factor II; FFP is fresh frozen plasma – see page 23

Factor II, VII, IX and X deficiency (Vitamin K-dependent coagulation factor deficiency

Severity range	Number of people in Scotland	Bleeding pattern	Inheritance	Treatment options
Mild–severe. Carriers usually have no symptoms.	< 6	Complex set of symptoms, including possible thrombosis.	Both parents need to pass on the gene.	Tranexamic acid, PCC (or possibly FFP) + Vitamin K1. Prophylaxis with oral Vitamin K1.

Factor V deficiency

Severity range	Number of people in Scotland	Bleeding pattern	Inheritance	Treatment options
Mild–severe. Carriers have no or mild symptoms, (Not to be confused with **Factor V Leiden**, which causes thrombosis.)	20	Nosebleeds, bleeding after operation or dental surgery, heavy periods.	Both parents need to pass on the gene	FFP, tranexamic acid.

Combined Factor V and Factor VIII deficiency

Severity range	Number of people in Scotland	Bleeding pattern	Inheritance	Treatment options
Mild–moderate.	< 6	Nosebleeds, easy bruising, gum bleeds.	Both parents need to pass on the gene.	Tranexamic acid, FFP and either recombinant Factor VIII or desmopressin.

Factor VII deficiency

Severity range	Number of people in Scotland	Bleeding pattern	Inheritance	Treatment options
Mild–severe. Carriers have no or mild symptoms.	196	Nosebleeds, gum bleeds, easy bruising, bleeding post-operation or dental surgery, heavy periods.	Both parents need to pass on the gene.	Recombinant Factor VIIa, tranexamic acid.

Factor X deficiency (Stuart-Prower factor deficiency)

Severity range	Number of people in Scotland	Bleeding pattern	Inheritance	Treatment options
Mild–severe. Carriers rarely affected.	43	Nosebleeds. In severe cases: after cutting umbilical cord; joint, head, stomach, surface and soft tissue bleeds; heavy periods.	Both parents need to pass on the gene.	PCC or Factor X (or FFP if neither available), tranexamic acid. Prophylaxis is possible.

Factor XI deficiency (haemophilia C)

Severity range	Number of people in Scotland	Bleeding pattern	Inheritance	Treatment options
Mild–severe. Carriers often only mildly affected.	200	Usually only after trauma or surgery (including childbirth).	Either one or both of the parents need to pass on gene.	Factor XI or tranexamic acid, (or FFP + possibly tranexamic acid). No recommendation for prophylaxis.

Sometimes known as haemophilia C, factor XI deficiency is the third most common inherited bleeding disorder. It is particularly common amongst Ashkenazi Jews (those traditionally from Eastern Europe), where 9% of the population are carriers and mildly affected. Unlike haemophilia A and B, the natural level of factor XI in the blood does not predict your tendency to bleed that well.

Factor XIII deficiency

Severity range	Number of people in Scotland	Bleeding pattern	Inheritance	Treatment options
Mild–severe. Carriers usually have no symptoms..	< 6	Bleeding into joints and soft tissue, head bleeds, poor wound healing, early loss of pregnancy, bruising, bleeding after cutting umbilical cord.	Both parents need to pass on the gene	Tranexamic acid, recombinant (or sometimes plasma-derived) Factor XIII. Prophylaxis is common.

Combined diagnoses, miscellaneous and unclassified bleeding disorders

Severity range	Number of people in Scotland	Bleeding pattern	Inheritance	Treatment options
Mild–severe. Some people are known to have more than one bleeding disorder, others have complex or uncertain diagnoses, but do have symptoms.	84	Varies.	Complex, depending on type of combination	Depends on diagnosis. FFP and/or platelets may be used in an emergency or if there is no diagnosis.

Von Willebrands

Von Willebrands (sometimes called **von Willebrands disorder**, **von Willebrands disease** or **VWD**) is a relatively common bleeding disorder, thought to affect up to 1% of the world's population. Relatively few are actually diagnosed — 1043 out of a population of 5.35 million, which is 0.02% of people in Scotland.

For clotting factor VIII (see haemophilia A) to do its job of helping blood to clot, it needs the assistance of the von Willebrand factor (VWF). VWF also helps platelets clump together to form a clot. Von Willebrands is caused when there is a shortage of VWF in the blood or when the VWF that is present doesn't work properly.

Most people who are diagnosed with von Willebrands usually experience milder symptoms of bleeding. Unlike haemophilia, internal bleeds are rare but bleeding at the **surface** of the body, such as bruising, nose bleeds, mucosal bleeding from cuts, and heavy periods in women are common features (see p22). Bleeding after injury, childbirth, surgery and dental extractions (and other minor surgery) is also common. There are three main types of von Willebrands numbered from 1 to 3. Within Type 2 there are a further four sub-types: 2A, 2B, 2M and 2N. Type 2N is also known as **Normandy**.

Type	No. diagnosed in Scotland	Sub-type	Severe	Moderate	Mild	
1	376		•	•	•	Usually mild
2	24	2A	•			
	13	2B	•	•		
	27	2M	•	•		
	< 6	2N		•		Resembles mild haemophilia A
	16	Unknown				
3	13		•			
Unknown	665					Carriers may have mild symptoms.

Most types of von Willebrands are inherited in a **dominant** way, so if you have a subtype of von Willebrands that is inherited in this way, you only need to acquire one gene from one parent to be fully affected (see p19). Therefore if you are a parent with von Willebrands, then on average half of your children may be affected. Exceptions are with Type 2N and Type 3 von Willebrands, which are inherited in a **recessive** way, so both parents need to be carriers. **Type 3** is also generally the most severe form of von Willebrands, and people may have **joint and muscle bleeds** as well as the pattern of bleeding described above. It is also the rarest — generally only up to 5% of people diagnosed with von Willebrands have type 3. Females are just as likely to be affected as males and in fact more women than men are diagnosed because of heavy periods or bleeding after childbirth (see pages 34–35).

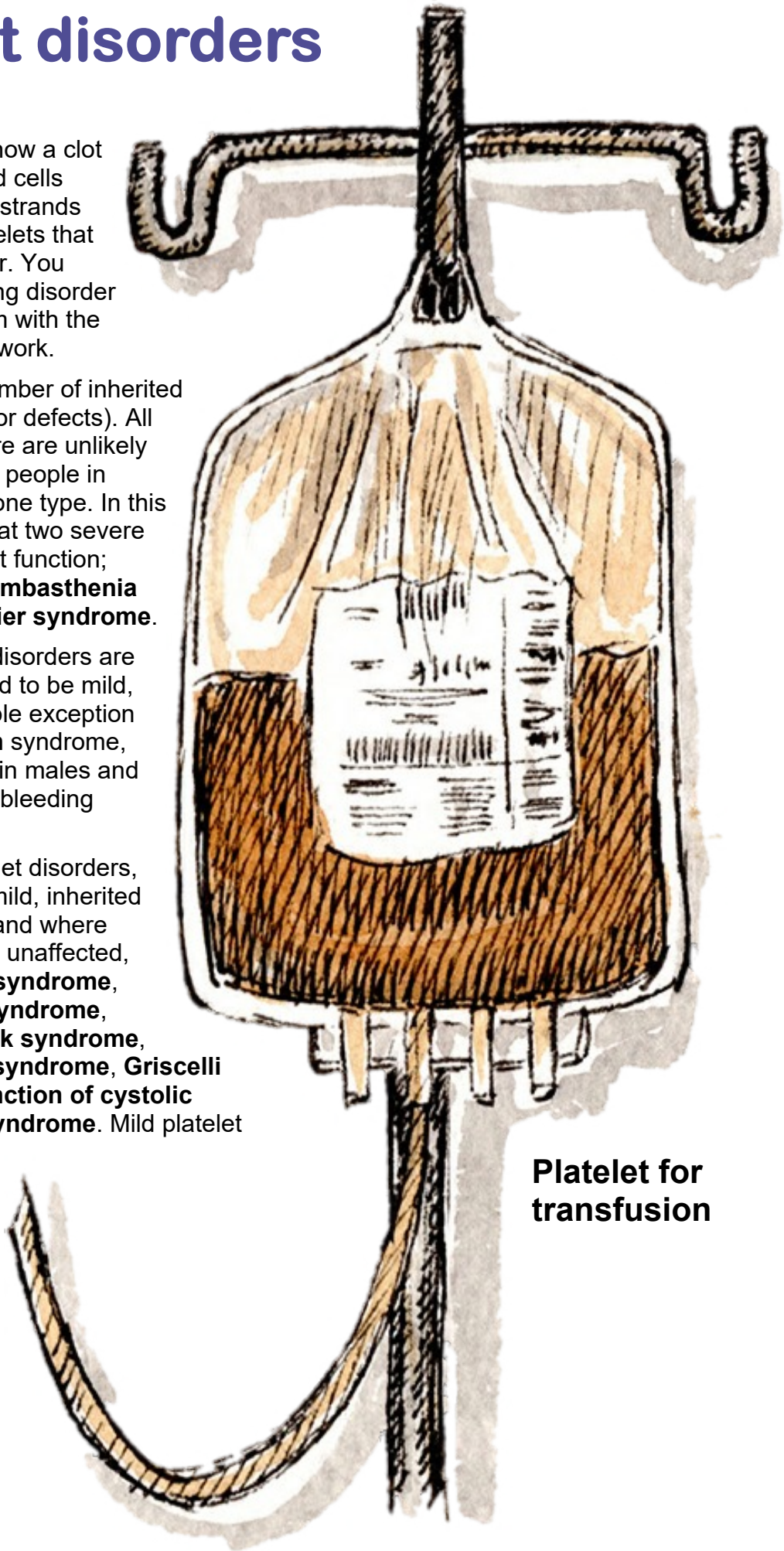
Platelet disorders

Page 5 described how a clot is made up of blood cells tangled up in fibrin strands and a mass of platelets that have stuck together. You may have a bleeding disorder if there is a problem with the way your platelets work.

There is a huge number of inherited platelet disorders (or defects). All are very rare – there are unlikely to be more than 12 people in Scotland with any one type. In this handbook we look at two severe disorders of platelet function; **Glanzmann's thrombasthenia** and **Bernard-Soulier syndrome**.

The other platelet disorders are normally considered to be mild, although one notable exception is **Wiskott–Aldrich** syndrome, which is only seen in males and has additional non-bleeding symptoms.

Other named platelet disorders, which are usually mild, inherited from both parents and where carriers are usually unaffected, are: **grey platelet syndrome**, **Quebec platelet syndrome**, **Hermansky-Pudlak syndrome**, **Chediak-Higashi syndrome**, **Griscelli syndrome**, **dysfunction of cystolic enzymes**, **Scott syndrome**. Mild platelet disorders which are inherited from only one affected parent are **platelet-type VWD** and **the non-muscle myosin heavy chain gene abnormalities**.



Platelet for transfusion

Glanzmann's Thrombasthenia

Severity range	Number of people in Scotland	Bleeding pattern	Inheritance	Treatment options
Mild–severe. Carriers have no symptoms	12	Easy bruising, nose bleeds, bleeding from gums, heavy periods; bleeding after childbirth, surgery, circumcision, or dental work.	Both parents need to pass on the gene.	Platelet transfusion, recombinant Factor VIIa, or both.

Bernard-Soulier syndrome

Severity range	Number of people in Scotland	Bleeding pattern	Inheritance	Treatment options
Mild–severe. Carriers have no or mild symptoms.	7	Most common symptoms are frequent nosebleeds, bleeding from gums, and easy bruising. Also after major trauma or surgery, and (less of a problem) dental extraction.	Usually both parents need to pass on the gene.	First approach is tranexamic acid and desmospressin. Major bleeds with platelet transfusion and/or recombinant Factor VIIa .

Inhibitors

You get an inhibitor when your immune system starts rejecting the clotting factor that is being used to treat bleeds. Treatment with that clotting factor no longer works, and other ways of stopping bleeding have to be used.

Inhibitors can happen in any clotting factor deficiency, including von Willebrands. However, they are most commonly experienced by people with severe haemophilia A – around 30% have, or have previously had, an inhibitor to Factor VIII. It is ten times less common in severe haemophilia B.

You can have varying amounts of inhibitor, and it may appear and rapidly disappear. A 'high titre' inhibitor is also the most difficult to get rid of. It is not usually possible to be on prophylaxis if you have an inhibitor (except to Factor VIII), so you should expect to have more bleeds.

Any of the following make you are more likely to get an inhibitor

severe haemophilia

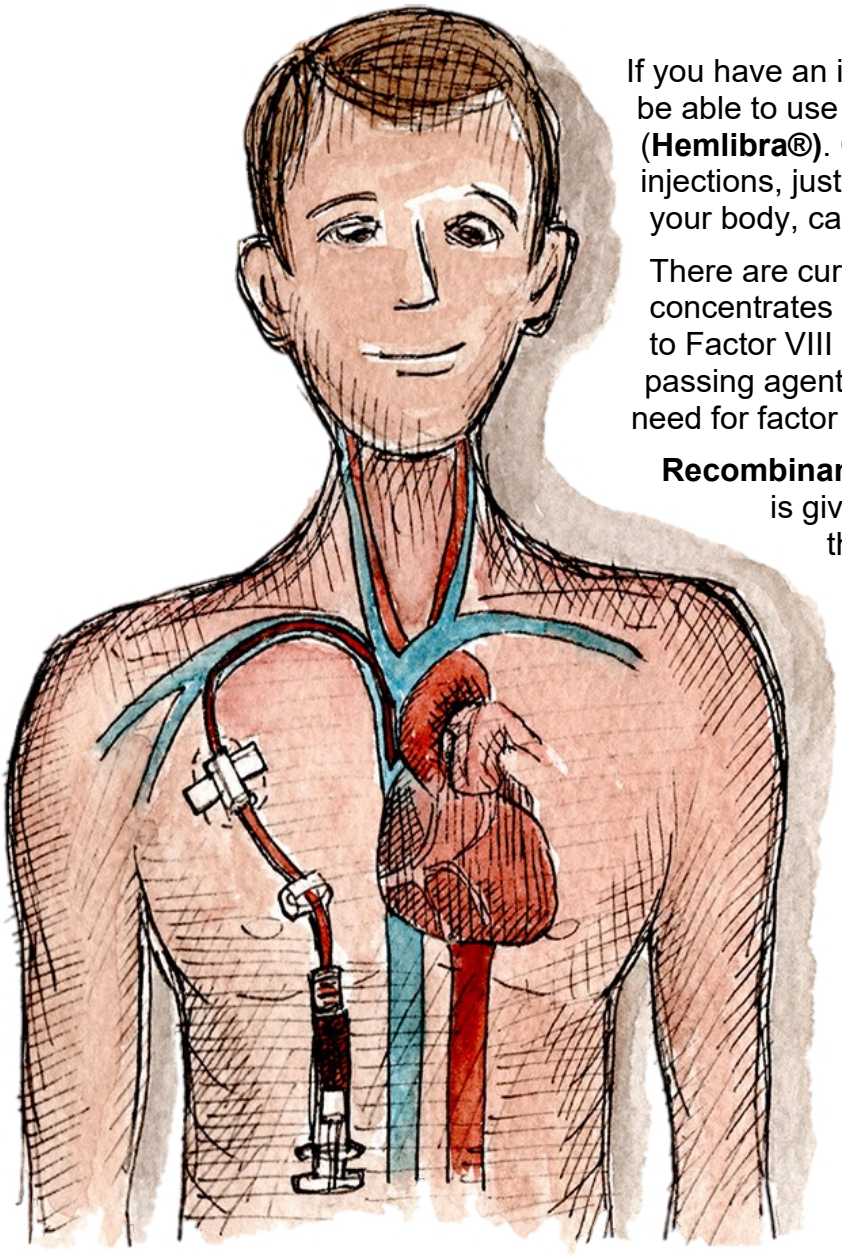
a family history of inhibitors

aged under 5,
or increasingly older than 60

recently started treatment

using a lot
of treatment

How to prevent or manage bleeds with an inhibitor to Factor VIII or IX



If you have an inhibitor to Factor VIII, you may be able to use a new treatment, Emicuzimab (**Hemlibra®)**. Giving yourself weekly injections, just under the skin in a fleshy part of your body, can help prevent bleeds.

There are currently two clotting factor concentrates that can be used as alternatives to Factor VIII or IX. These are known as 'by-passing agents', because they by-pass the need for factor VIII or IX:

Recombinant Factor VIIa (or **NovoSeven®**) is given by a single injection or up to three injections, two hours apart.

FEIBA, which stands for **Factor Eight Inhibitor Bypassing Activity**, is a type of blood product known as an **activated Prothrombin Complex Concentrate** (**aPCC**). It is the only aPCC currently available for use in Scotland. It is given as a single infusion followed by more after eight to twelve hours if necessary.

It may be possible to simply use a higher dose of Factor VIII or IX than you used before you had an inhibitor, or if you have mild haemophilia A, to switch to desmopressin. **Tranexamic acid** can be used in addition, especially for surface bleeds, but is not with FEIBA.

A port-a-cath is often used for if you need frequent injections to remove an inhibitor, or for regular prophylaxis (see p 23)

How to get rid of an inhibitor to Factor VIII

The first line of attack if you have severe haemophilia A and an inhibitor is a tactic known as **immune tolerance therapy**. By infusing large amounts of Factor VIII daily for up to 18 months you try to 'drown out' your inhibitor by overloading it with the clotting factor that it is neutralising. If this is not successful, then you can try a higher dose, switch to plasma-derived clotting factor, or use **rituximab**, which is a drug that reduces the activity of your immune system.

Treatments explained

Good treatments to stop or prevent bleeding now exist for most bleeding disorders. Some are specific to a particular type of bleeding disorder – for example, Factor IX concentrate for haemophilia B. Others can have widespread use, such as tranexamic acid for any bleeding disorder. A **concentrate** is a powder that you dissolve and then inject into one of your veins. Sterile water provided from the manufacturer or the haemophilia centre, or sodium chloride (salt water) are the commonest ways to dissolve the powder.

	Haemophilia A	Haemophilia B	Von Willebrands	Other clotting factor deficiencies	Platelet disorders
Recombinant FVIII	✓				
Recombinant FIX		✓			
Plasma FVIII with vWF			✓*		
Recombinant FVIIa	✓*	✓*		✓	✓
aPPC	✓*	✓*			
Emicuzimab	✓*				
Other clotting factors				✓	
Platelets					✓
Fibrin glue	✓	✓	✓	✓	✓
Tranexamic acid	✓	✓	✓	✓	✓
Desmopressin	✓		✓	✓	✓
Fresh Frozen Plasma				✓	
Cryoprecipitate				✓	

**May be used if you have an inhibitor to Factor VIII or IX*

Factor VIII and IX

Factors VIII and IX were first extracted from human blood in the 1960s and they revolutionised treatment for bleeding in people with haemophilia A and B, even allowing for patients to treat themselves at home. These concentrates were in widespread use from the 1970s until Scotland started to replace them with genetically-engineered forms (known as **recombinant products**) in the mid 1990s because they have a theoretically zero risk of spreading any infection.

How often to inject prophylaxis treatment		
	Conventional recombinant/plasma	Longer-acting
Factor VIII	Normally every 2 or 3 days	Every 3–5 days
Factor IX	Normally every 3 or 4 days	Every 7–14 days

Although ‘recombinant’ is the treatment of choice for severe haemophilia A and B, some people continue to prefer concentrates that have been made from **plasma** (the liquid part of human blood). If you have von Willebrands you may need to use a plasma-derived Factor VIII that also contains a good amount of von Willebrand factor. This type of Factor VIII can also be useful if you have haemophilia A with an inhibitor. You can use all these treatments **on demand** (when bleeding), or for **prophylaxis**.

The latest development in clotting factor treatment is the preparation of **longer-acting** concentrates. If on prophylaxis you infuse these less often than conventional Factor VIII and IX concentrates.

Treatments for inhibitors to Factor VIII and IX: NovoSeven, FEIBA and HEMLIBRA

Recombinant Factor VIIa treatment (the ‘a’ stands for ‘activated’) is better known by its brand name, **NovoSeven®**. It is mainly used to treat bleeds or as pre-surgery prophylaxis in people with an **inhibitor to Factor VIII or IX**. It is also licensed for **factor VII deficiency** and the platelet disorder, **Glanzmann's thrombasthenia**.

FEIBA (pronounced “Feeba”) is a concentrate made from plasma that is mainly used if you have an **inhibitor to Factor VIII** (on demand or prophylaxis, see p13).

HEMLIBRA® is a type of medicine known as a therapeutic antibody. It can bridge activated Factor IX and Factor X in your blood to replace the function of missing activated Factor VIII in people who have an **inhibitor to Factor VIII**.

Other clotting factor concentrates

A range of other single clotting factor concentrates to treat rarer conditions is available, but their availability can vary over time. Currently two are recombinant – for **factor VII and XIII deficiency**, and the rest are made from plasma from non-UK donors – for **Fibrinogen and factor VII, X, XI and XIII deficiencies**. Concentrates containing four clotting factors exist (II, VII, IX and X) and are preferred if you have **factor II or X deficiency** (see aPCC, p12).

Desmopressin

Desmopressin (also known as **DDAVP**) is a synthetic hormone that treats bleeds in **mild** (and sometimes **moderate**) **haemophilia A** and most forms of **von Willebrands (types 1, 2A, 2M and 2N)**. Because it is not made from blood, it has always been safe from any blood-borne infections. But it does cause your body to retain water and can make your skin temporarily red and hot (flushing). It can reduce your blood pressure and even cause thrombosis (clotting). You will be given a trial dose to check how well it works and see if there are any side-effects.

Platelets

Platelet transfusions may be used if you have a **severe platelet disorder**. They are obtained from voluntary donors.

Fresh Frozen Plasma (FFP)

FFP, from voluntary donors, contains all the clotting factors you need and is used when one of them is not manufactured or available, or when your bleeding disorder has not been diagnosed. It is treated to greatly reduce the risk of infection.

Cryoprecipitate

Cryoprecipitate is made from plasma and contains fibrinogen, factor VIII, factor XIII and the von Willebrand factor. It is used if Fibrinogen or Factor XIII concentrates aren't available.

Tranexamic acid

Tranexamic acid helps prevent clots break up naturally. It can be taken as a tablet, a mouthwash (before dental work) or an infusion. It is often used for mouth bleeds or before dental surgery, and by women with heavy periods. Epsilon-Aminocaproic acid (EACA) is a similar drug.

Fibrin glue

As its name suggests, fibrin glue helps clots form and is used by dentists and surgeons and other people treating bleeds.

How to stay well with a bleeding disorder

In most cases it is now possible to lead a full and active life with a bleeding disorder, and you can expect to live as long as someone without one. Here are five top tips to help you stay well, but you will discover more as you learn to live with your bleeding disorder.

Tip 1: Treat bleeds early

Learn how to spot the signs of an internal bleed. You may have bumped into something hard and begun to see bruising on top of the normal pain of injury, or you may have known that your child had fallen over. People with a severe bleeding disorder can also bleed without any noticeable injury. This is known as a **spontaneous bleed** and may be less visible and the pain may be delayed.

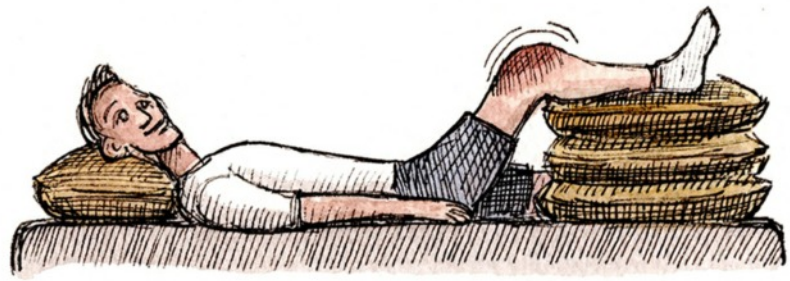
If you have treatment at home or carry it with you then you can treat yourself, but you can also seek advice and/or treatment from your regular haemophilia centre during clinic open hours or by contacting the on-call doctor. Your Centre should give you this information. If it is an evening or weekend, go to your hospital's Emergency Department and on the way try to contact their on-call haematology team to help ensure the quickest possible treatment when you arrive. The principles of **R.I.C.E.** are good to follow, especially if you can't access treatment straight away.

Rest – use a sling or pillows, don't move the limb or walk on it.

Ice – wrap ice in a damp towel and apply for five minutes, remove for ten, then repeat.

Compression – use a stretchy bandage or elastic stocking (but not if it hurts a nerve).

Elevation – raise the area that is bleeding above the heart.



Tip 2: Use your prophylaxis treatment to help prevent bleeds

People with **severe haemophilia A and B**, and also sometimes **other bleeding disorders**, may treat themselves regularly, even if there is no sign of bleeding. This is known as **prophylaxis** – it helps protect the body from spontaneous bleeds and bleeding from smaller injuries.

Tip 3: Do the right sort of exercise to help prevent bleeds

It's easy to think that any sort of exercise could cause a bleed, but in fact the opposite is true. Sport and most forms of exercise help build up muscle strength, which helps prevent spontaneous bleeds and joint damage. But it's important to enjoy the right sort of sport: swimming, badminton, cycling and walking are generally safe, whilst contact sports such as rugby or boxing are not recommended. Take advice and come to your own decision about which sports in between these two extremes are appropriate for you.

The National Hemophilia Federation in the USA has produced a detailed analysis of many types of sport and exercise. The more common types are listed in the box.

Lowest risk	Low to moderate risk	Intermediate risk	Highest risk
● Elliptical machine training ● Golf ● Exercise bike ● Swimming (not diving) ● Walking	● Circuit/strength training ● Rowing ● Treadmill running	● Athletics ● Cycling ● Dance (some) ● Zumba ● Yoga ● Gymnastics ● High intensity training ● Football ● Running ● Tennis	● Bouncy castle ● Boxing ● Wrestling ● Martial arts (some) ● BMX racing ● Hockey ● Lacrosse ● Weight-lifting ● Rugby

Tip 4: Don't take aspirin and other non-steroidal anti-inflammatory drugs (NSAIDS)

These drugs are often used to control pain or a high temperature, and **aspirin**, **ibuprofen** and other forms of NSAIDs are available over-the-counter. They stop platelets working (and therefore hinder clotting) and make stomach bleeds more likely. They, or any cold or 'flu remedy that contains them, should not be taken without medical advice.

Tip 5: Tell all your healthcare professionals and dentists about your bleeding disorder

It's vital that only the right drugs are prescribed and the risk of bleeding from any procedure (e.g. dentistry and major or minor surgery) is well managed.

Lewis is 11 and likes football and swimming	Andrew is 48 and a fork-lift truck driver
Lewis is a typical active 11-year-old boy with severe haemophilia A. He knows that if he doesn't take his Factor VIII regularly he puts himself at greater risk of having a bleed, which means he can't enjoy sports for a few days. He treats himself before school on Mondays and Thursdays so that he has maximum protection for all his activities.	Andrew has moderate haemophilia B. His warehouse has a good safety record, but he has learnt to keep his Factor IX with him at work in case he knocks himself. If he does have an injury, he can usually treat himself straightaway and do some office work for the rest of the day rather than have to go to hospital.

Tip 6: Take good care of your teeth and gums

It is important for you to look after your mouth and teeth as you can be affected by both dental decay (caries) and bleeding gums (periodontal disease). These conditions affect people who do not have bleeding disorders. It has been shown that with good oral care and support from your dentist that the incidence of bleeding from the gums can be almost eliminated.

You should register with a dentist and let the doctors and nurses in the haemophilia centre the name and address of your dentist. Children should be registered with a dentist before they are 6 months old and attend for regular examinations. Your dentist will concentrate on preventing dental problems and discuss foods which are likely to cause problems. As you get older the frequency of visits may alter and you may also be asked to see the dentist who works with your haemophilia centre occasionally as you may need to be referred to them if you every require a tooth removed.

Your dentist will need to discuss any treatment plans with your haemophilia doctor and the timing may need to be adjusted to allow for treatment from the haemophilia centre. It should be possible for your dentist to provide the same comprehensive care as any other patient. A few patients may need some of their treatment carried out in a hospital so that you can be monitored by the haemophilia centre. This will be discussed with you as required.

If you look after your teeth they will last a lifetime and if you follow the advice on a healthy diet, regular tooth-brushing and visit your dentist this will reduce the need for dental care.

If you have problems registering with a dentist please discuss this at your next visit to the haemophilia centre and they will be able to help.

How bleeding disorders are inherited

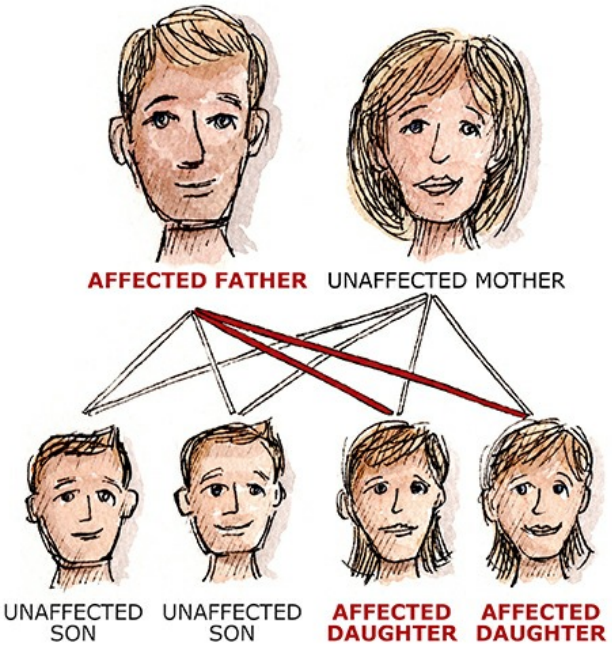
Virtually all the cells in our bodies contain a common set of DNA that carries the instructions for everything that the body does. Within this DNA are genes to make clotting factors. If any gene is missing or faulty, your body can't make the corresponding clotting factor or if it can, the clotting factor won't work properly.

Since you inherit half your DNA from your father and half from your mother, if either of them are affected by a genetic bleeding disorder then you might be also. If you have inherited a single bleeding disorder gene from one parent but don't show any symptoms you are known as a **carrier**, because you can still pass the gene onto your children.

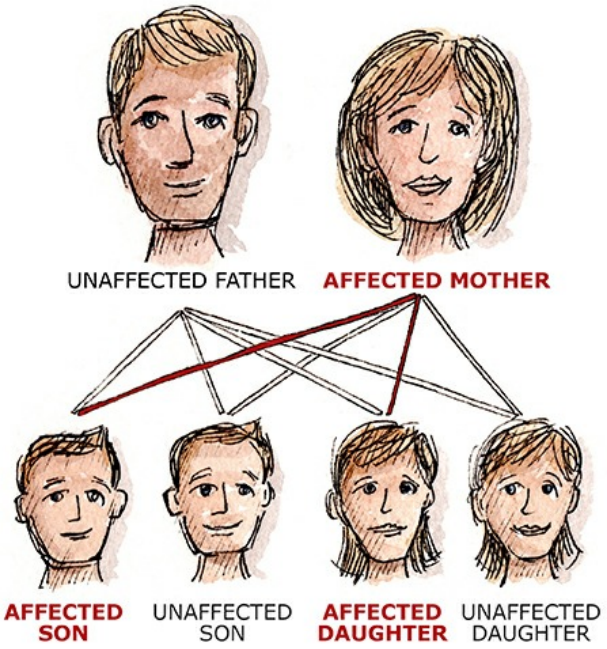
If you have a parent or other close family member who has been diagnosed with a bleeding disorder, you can ask your local haemophilia centre or GP for genetic counselling and testing. If you find out that you also have the gene it will help you understand the possibility of having a child with a bleeding disorder.

Haemophilia A and B

The way the haemophilia A or B gene is passed on depends on the gender you were assigned at birth. Either parent can pass on the gene: the diagrams show the two main ways this can happen. Women and girls with one haemophilia gene often have minor bleeding symptoms and are all carriers. Haemophilia Scotland offers support to all such females, regardless of their clotting factor level, as carrying the gene can affect their decisions.



His sons cannot inherit haemophilia, but all of his daughters are affected.



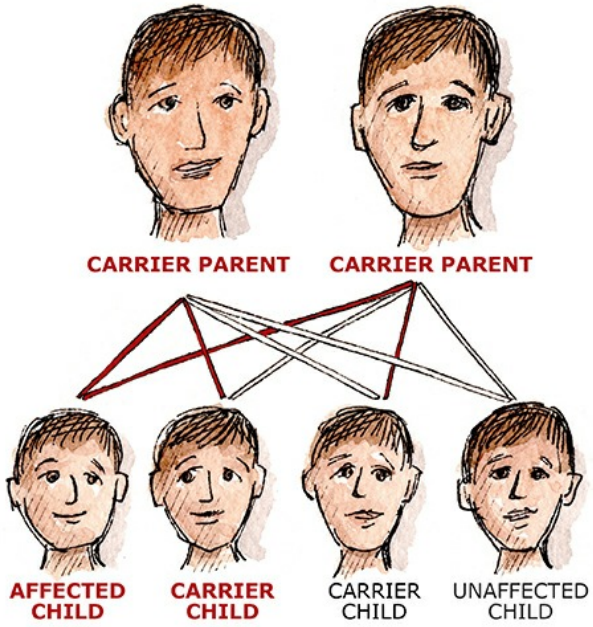
Her sons have a 50:50 chance of having haemophilia, and her daughters have a 50:50 chance of being affected.

Von Willebrands and other inherited bleeding disorders

The rest of the bleeding disorders featured in this handbook are inherited in a way that does not depend on your sex or the sex of your parents. Certain rare conditions can be complex, and sometimes you can be classed as a carrier and still have symptoms, but generally they are inherited in one of two ways, known as **recessive** or **dominant**.

Recessive inheritance (from both parents)

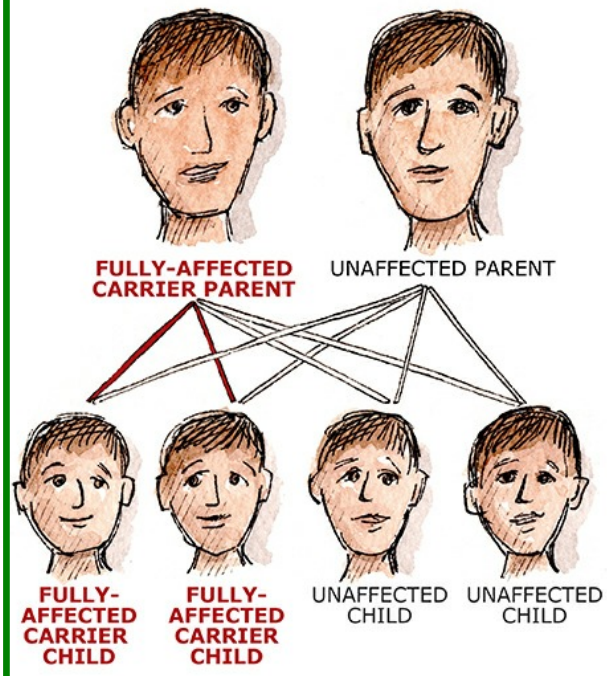
- Types 2N and 3 von Willebrands
- All single clotting factor deficiencies (apart from some of the fibrinogen defects)
- Most platelet deficiencies



Both parents need to be carriers. Their children have a 1:4 chance of being fully affected and a 50:50 chance of being carriers.

Dominant inheritance (from one parent)

- Types 1 and 2A, 2B and 2M von Willebrands
- Dysfibrinogenaemia



Only one parent needs to be affected. Their children have a 50:50 chance of being affected.

Inheriting bleeding disorders when both parents are affected

It is very unusual for both parents to carry the same bleeding disorder gene. If they do, then there is a greater chance of having affected children than shown in the above diagrams. This is most likely to happen when close relatives, such as first cousins, have children together, and is why type 3 von Willebrands is relatively more frequent in cultures where this practice is common.

Bleeding disorders that are not inherited

In very rare circumstances, particularly in old age or pregnancy, it is possible to develop what is known as '**acquired haemophilia'** or '**acquired von Willebrand Syndrome**'. This happens when your body's immune system starts to reject a clotting factor that helps your blood to clot. These conditions are not genetic.

There are many other non-inherited medical conditions that cause bleeding such as **immune (idiopathic) thrombocytopenic purpura (ITP**), some liver conditions and some types of cancer and cancer treatments.

Looking after a child with a bleeding disorder

As soon as your child's bleeding disorder is diagnosed, you should be put in contact with your local haemophilia centre. Staff will arrange regular review appointments and give you advice on how to spot and treat a bleed, and what to do in an emergency.

How to spot a bleed


Photo by rawpixel.com from Pexels

The type and severity of bleeding that your child experiences will depend on what bleeding disorder he or she has. Bleeding and bruising from cuts are common in von Willebrands and are easy to spot. But with more severe bleeding disorders (especially haemophilia) people bleed into their joints or muscles without even injuring themselves. In time, an alarming darkly-coloured bump under the skin may form, which may cause a joint (such as an ankle, knee or elbow) to swell considerably. Before you can see any sign, your child may be in a lot of pain. They may not tell you that they are hurting, because they've learned that a bleed means stopping whatever they are doing. Look out for a difference in the way they use their left and right legs, or arms and hands. They might describe a heat feeling or tingling sensation.

Bleeding inside the skull is always an emergency that requires immediate hospital treatment. Your child may have a severe headache, feel or be sick, move oddly and could

Allegations of non-accidental injury

Bruising is common across a wide range of bleeding disorders. As well as being alarming for your child, you might be accused of child abuse. General information such as this handbook, or a letter from your doctor, will help if you're investigated by social services.

Leaving your child in others' hands

Childminders, nurseries and schools can be reluctant to look after a child with a bleeding disorder because of an irrational but understandable fear of the consequences of a bleed. Educational materials such as this handbook and a visit from your haemophilia nurse can help put their minds at ease. If they still refuse to take charge of your child they risk falling foul of disability discrimination regulations within the UK-wide Equalities Act 2010.

Being over-protective?

A common but understandable reaction to caring for a child with a bleeding disorder is to be over-protective, taking extreme steps to avoid any injury that might cause a bleed. This can mean the child misses out on playing with friends and enjoying healthy activities, which would actually make joints stronger and helps prevent bleeds. Wearing a helmet and other protection is rarely recommended, unless on a bike, scooter etc., but your haemophilia centre's medical team can always advise.

When the treatment is working well, Haemophilia Scotland recommends you aim for your child to enjoy most activities that their friends take part in, and to lead a healthy and active life (see p 17).

Encouraging independence

Children can learn to help prepare their treatment and, in time, self-inject it at a surprisingly early age. This is often the first step in learning to become more independent in managing their own bleeding disorder. As they grow older, they will become more expert in their illness and better at spotting and managing a bleed. On leaving home and having transitioned from a children's to an adult's haemophilia centre, the medical team will further support them to become independent and increase their confidence.


Photo by Teemu R from Pexels

How to spot a joint or muscle bleed

With a joint bleed, it may not be obvious at the start, so look out for the joint becoming hot, swollen and tender, or the person saying they feel tingling, pain or stiffness. If they can't, or don't want to, talk about what they are feeling, then look out for visual signs of discomfort and different-sized arms or legs, difficulty in movement, or them avoiding using that arm or leg.

Muscle bleeds are most often in the calves, forearms, bottom, thighs and groin. They also cause swelling and pain.

How to spot a stomach/gastrointestinal or urinary tract bleed

- Bloody or black tar-like poo
- Red or brown urine

How to spot a head bleed

- severe headache
- stiff neck
- vomiting
- change in mental state, such as confusion
- speaking difficulties, such as slurred speech
- changes in vision, such as double vision
- loss of co-ordination and balance
- paralysis of some or all of the facial muscles

Even if you haven't witnessed any injury, and can't see any obvious lumps or bruising, treat it as an emergency.

Issues for women

Traditionally women have done the lion's share of caring for people with a bleeding disorder. This is because they have generally been the main carer of children and may also have looked after a male partner with haemophilia. Increasingly there has been greater awareness that women can have symptoms of a bleeding disorder. Most often this will be as a carrier of haemophilia, possibly with mild bleeding symptoms, or living with von Willebrands.

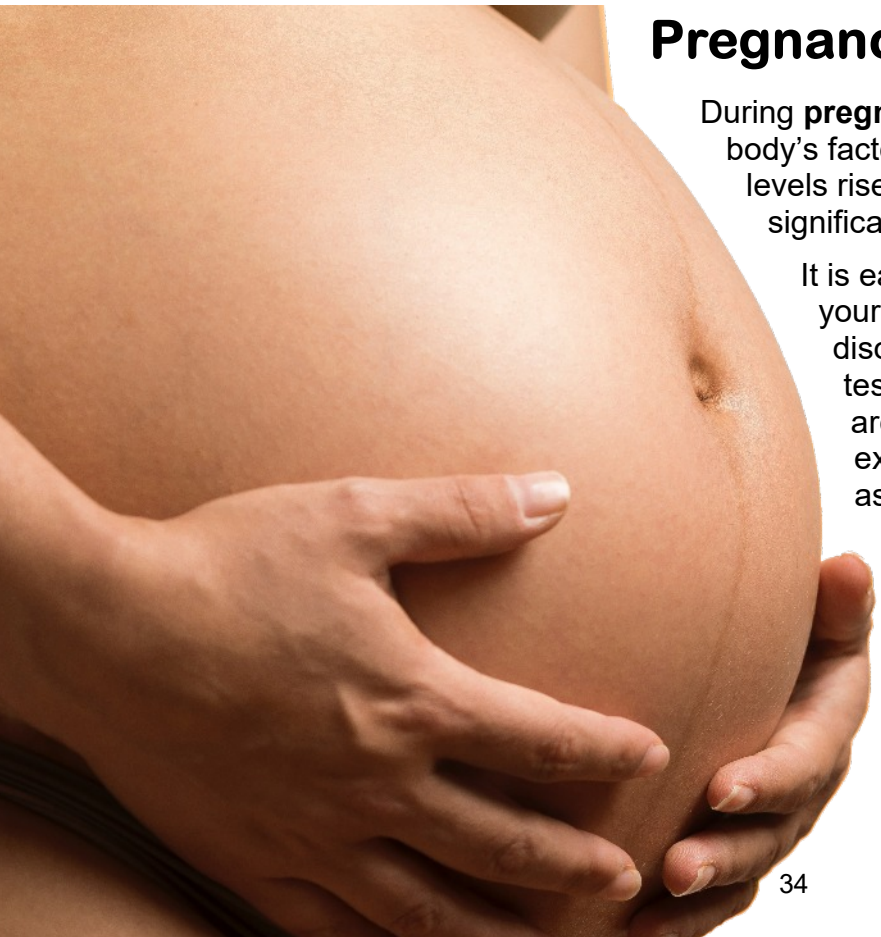
Amongst all bleeding disorders apart from haemophilia, women are more likely than men to be diagnosed because of heavy periods (menstrual bleeding) and bleeding after childbirth.

Pregnancy and childbirth require special management. The **Glasgow**, **Edinburgh and Aberdeen Centres** hold joint women's clinics with an obstetrician (childbirth consultant), and provide support to newborn babies with a bleeding disorder.

Heavy periods

Heavy menstrual bleeding is twice as likely to affect carriers of haemophilia as all other women, and is extremely common with von Willebrands (even if Type 1, which is the mildest type). You would expect to lose a greater amount of blood each month and have longer periods, with more pain. In mid-cycle there can be further bleeding and pain. The greater loss of blood and therefore iron can cause long-term anaemia.

For decades, heavy menstrual bleeding (whatever its cause) has been managed with **tranexamic acid** (see p15). Women who are affected by haemophilia A with low Factor VIII levels, or mild-to-moderate Type 1 von Willebrands, can also use **desmopressin**. The impact of periods can be greatly reduced by going on the (combined) contraceptive pill or having a Mirena coil fitted. But despite these measures it is still possible that clotting factor treatment will be required.



Pregnancy

During **pregnancy** the good news is that your body's factor VIII (and von Willebrand factor) levels rise (although factor IX doesn't significantly change).

It is easier to plan the birth if you know if your baby is going to have a bleeding disorder. Its sex can be found out by testing your blood or by ultrasound. If you are a carrier of haemophilia and are expecting a boy, then procedures such as chorionic villus sampling (CVS) can tell the difference between an affected and unaffected male, but there is a 1-2% risk of miscarriage. Increasingly, genetic laboratories are able to detect small amounts of your baby's DNA in your own blood. This is a safer way of finding out your baby's sex before birth.

Childbirth

During labour and delivery, both mother and baby may need extra care. If your factor levels are still low at the time of delivery, expect to deliver the baby at a hospital with specialist haemophilia care, using a birth plan that has been agreed with your haemophilia team. As well as being watchful for your baby showing signs of bleeding inside its skull, you might need treatment after birth.

Caesarean sections are becoming increasingly common, especially when babies have severe haemophilia. But they are not considered completely essential. If you have a natural childbirth, there are greater risks to your baby if ventouse or (to a lesser extent) forceps are used, or the labour is prolonged, scalp electrodes have been used on the baby or its blood has been sampled. After birth, a sample of umbilical cord blood will be taken to help you learn whether your new-born could have haemophilia, but any diagnosis will need to be reviewed when the baby is older.

Fiona's story

When I was born in 1981, I don't think anyone in my family had heard of haemophilia, let alone think I might have to face up to it. This was all to change in 2005 when I had my first son. I bled quite a lot after giving birth, and they told me I had needed a blood transfusion (although I can't really remember). Lewis was a very healthy baby, but he did seem quite bruised and, thinking back, he bled a lot from where the umbilical cord was cut and when he had his heel prick test. He started crawling at nine months and it was then that I noticed he suffered quite bad bruising whenever he bumped into things. In fact I had a visit from social services, but mercifully the social worker knew something about bleeding disorders and said that the pattern of bruising could well be due to something like haemophilia.

I got this checked out and, lo and behold, he had severe haemophilia A! The haemophilia centre asked if I would like to be tested, because I *might* be a carrier – although it was possible I wasn't and Lewis just had an out-of-the-blue altered factor VIII gene. It turned out I *was* a carrier, and in fact had fairly low factor VIII in my blood, which explains why I bled after giving birth and have always had bad periods. I just thought that was normal for me and hadn't had it checked out. I'm not with Lewis's father any more, but if I did get pregnant by someone else, I know to expect there's an evens chance that if it's a boy, he'll have haemophilia. It's the same odds that a girl will be a carrier, and perhaps grow up with symptoms like me. I've come to terms with my haemophilia and I just get on with what's really a pretty ordinary life. I'm sure my children will, too.

It's easy to be worried by all this information, but it's worth remembering that most women with a bleeding disorder learn to live with their condition and are able to lead normal lives. And very many babies, even with severe haemophilia, are safely born without any precautions being taken because there hadn't been any family history of a bleeding disorder.

The practical side of living with a bleeding disorder

Self-treatment and home delivery

People with the most common types of severe bleeding disorders are able to keep treatment at home in case of a bleed and also for prophylaxis. A nurse can show you how to dissolve the concentrate in a vial of sterile water and use the manufacturer's specially designed syringe and needle to inject the treatment into one of your veins. Some people needing very regular treatment, especially children, often have a **port-a-cath** surgically fitted for easy access to a vein in the chest.

Home-delivery of treatment benefits over 200 patients in Scotland. Check the vials' expiry dates for how long you can keep them and note the temperature they need to be stored at. You will also need to keep records of the dates of and how much treatment you have injected.

Food and drugs to avoid

Your haemophilia centre can always advise about what food and drugs promote bleeding. Drugs that should not be taken (or only used in consultation with your haematologist) include non-steroidal anti-inflammatory drugs (NSAIDs, see p16) and certain anti-depressives.

Some herbal drugs are also not recommended. These include:

- Ginkgo biloba
- Garlic in large amounts
- Ginger (not dried ginger)
- Ginseng (Asian)
- Feverfew
- Saw Palmetto
- Willow bark

Telling others about your condition

Many people in a new relationship worry about when and what to tell their partner. There are no hard-and-fast rules, but it is usually best to wait until they have got to know you and see how you lead a normal life. If you wait too long, however, you risk them feeling hurt about being kept in the dark. Sex is not usually a problem even if you have a severe bleeding disorder, but if you are living with a sexually transmitted infection from contaminated treatment used before safer alternatives became available, then using a condom will help prevent your partner from becoming infected.

New partners and friends will feel more reassured if they have information such as is found in this handbook.

The emotional side of bleeding disorders

Most people with a bleeding disorder who can access good treatment are able to lead normal lives and expect a near-normal life expectancy, particularly if their condition is mild or moderate. Emotionally, times are hardest when the bleeding disorder is first diagnosed, or someone has a bad bleed or develops an inhibitor, and at changes in life, such as moving away from the parental home. You'll often wish you weren't affected, and wonder why it happened to you and not another close family member. At times you may feel like ignoring the fact that you have a bleeding disorder at all. But by burying your head in the sand you neglect the major part you can play in managing your bleeding disorder and may suffer harm.

You will make mistakes, such as travelling without treatment, wishing you had treated a bleed that you thought didn't need treating, or forgetting your prophylaxis and as a result having a bleed. This is inevitable and it's important not to let it get you down but to learn from the experience.

Many people appreciate the opportunity to meet others in a similar position to share experiences and feelings, and learn from each other. Some go on a self-management course to learn skills to better cope with living with a bleeding disorder and get the most out of their haemophilia centre's services. Ask your centre or Haemophilia Scotland what groups and activities are available to you.

If you want to have a family you will have to consider the possibility of passing on the gene that gives rise to the bleeding disorder. It is possible to minimise the risk with techniques that use IVF to determine the sex of your baby, for example to ensure you don't have a boy with haemophilia. You also face the choice of investigating if your unborn baby is carrying a gene that would give rise to a bleeding disorder, and then deciding what to do with that information. Alternatively you may consider adoption or sperm donation to be the best way of having an unaffected child. These are not easy things to think through, and we recommend you seek expert help in coming to the best decision for you. Speak to your Centre about psychological support available to you.

Looking after someone with a bleeding disorder

Looking after someone else may turn into a lifelong commitment. As a carer you will become quite an expert in managing your partner's or family member's condition. Sometimes you'll wish they would do more for themselves, as you need time to focus on your own health and wellbeing. Alternatively, you might prefer to be more involved but feel you are not told everything. We encourage carers to attend clinical review appointments and access all the forms of support that are available.

Treatment and care in Scotland and beyond

There are six Haemophilia Centres situated in five Scottish cities and towns. Three of them are Comprehensive Care Centres (CCC) which means that they can offer the complete range of Haemophilia Services. You can choose which Haemophilia Centre you are registered at and you can change centre at any time. You may prefer to be registered at both a CCC in Glasgow or Edinburgh, and at a smaller centre outside these cities and local to you.

If you live a considerable distance from any of these centres, then your haemophilia care might be organised as a partnership between a Haemophilia Centre and your local hospital or GP.

Inverness
Raigmore Haemophilia Centre, Raigmore Hospital
IV2 3UJ
01463 704020

Aberdeen
NHS Grampian Haemophilia Centre, Aberdeen Royal Infirmary AB25 2ZN
01224 553357

Dundee
Dundee Haemophilia Centre, Ninewells Hospital
DD1 9SY
01382 632225

Edinburgh (CCC)
Haemophilia and Thrombosis Centre, Royal Infirmary of Edinburgh EH16 4SA
0131 242 1270

Glasgow Adults (Joint Glasgow CCC)
West of Scotland Adult Haemophilia and Thrombosis Centre, Glasgow Royal Infirmary
G4 0SF
0141 211 5127

Glasgow Children (Joint Glasgow CCC)
Paediatric Haemophilia CCC, Royal Hospital for Sick Children
G51 4TF
0141 452 4630

If you live in Scotland but are travelling in the rest of the UK, you will find treatment and care are readily accessible and there is a good network of CCCs and local Haemophilia Centres. Treatment is also free in countries that belong to the European Union and a few others (including Norway and Switzerland), but you are likely to rely on travel insurance to pay for any treatment elsewhere.

In an emergency, telephone your nearest hospital with an Emergency Department and ask to speak to the on-call haematologist. They can tell you which part of the hospital to meet them at.

Top tips for travellers

- Carry information about your bleeding disorder, including a letter from your haemophilia centre that describes what treatment you need, and consider wearing medical ID jewellery
- Arrange to take enough treatment with you to cover prophylaxis and bleeds
- Contact your airline/airport in advance if you need to take needles
- Keep your treatment with you as hand luggage, not in the aircraft's hold
- Make sure you have an up-to-date European Health Insurance Card (EHIC)

Take out travel insurance that includes cover for your bleeding disorder (which you must mention)

Carry with you a list of local haemophilia treatment centres (see the WFH website)

More information is available at www.haemophilia.scot/travel

This publication was co-produced by Haemophilia Scotland and the Scottish Inherited Bleeding Disorder Network. While every effort has been made to ensure the information it contains is accurate and up to date any changes in treatment or care must only be taken in consultation with your haemophilia centre in line with current guidance and protocols from the Scottish Inherited Bleeding Disorders Network and/or the United Kingdom Haemophilia Centre Doctors' Organisation (UKHCDO) or other relevant international bodies. We are grateful to all the healthcare professionals and patients who have contributed to this publication.

The content of this publication can also be found online at www.haemophilia.scot and www.sibdn.scot.nhs.uk.

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