

Girls living with bleeding disorders



**The
Haemophilia
Society**

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Introduction

Living with a bleeding disorder can be challenging to manage at times and may affect relationships, education, work and everyday life. It can also have a huge impact mentally, physically and financially. It can be emotional living with conditions that some people find hard to talk about.

As a girl or young woman with a bleeding disorder, it is helpful for you to understand your condition and the impact it may have. This booklet covers various bleeding symptoms, and it's important to note that bleeding disorders do not affect everyone in the same way.

Depending on your age and your particular bleeding disorder, you'll need different information at different times. The booklet is divided into sections on different topics so you can dip in and out of it. There's also a helpful list of medical words in the glossary at the back.

We hope the information in this booklet helps you in developing your knowledge. Other family members might find it useful too.

If you have any questions about anything you read in this booklet you can call the Haemophilia Society on **020 7939 0780** or email **info@haemophilia.org.uk**

Section 1 - Bleeding disorders

What are bleeding disorders?

Bleeding disorders are conditions where the blood doesn't clot properly. Most are inherited and are caused by either a blood clotting factor or platelets not working correctly or being reduced or even missing altogether.

Clotting factors are proteins in the blood that control bleeding. They are written in Roman numerals, and each related bleeding disorder is named after the clotting factor that is lacking. There are many different types of bleeding disorders because many other proteins (factors) are involved in normal blood clotting.

These are bleeding disorders caused by problems with clotting factor deficiency:

Factor I (one) [fibrinogen]	Factor II (two) [prothrombin]	Factor V (five)
Factor VII (seven)	Factor VIII (eight)	Factor IX (nine)
Factor X (ten)	Factor XI (eleven)	Factor XIII (thirteen)
von Willebrand factor (VWF)		

Your doctor may call your disorder a 'deficiency'. You may hear your bleeding disorder called factor XI deficiency, for example.

Platelets are small cells in the blood that clump together to start the process of forming a clot. When this doesn't work properly you have a 'platelet function disorder'.

Haemophilia

Haemophilia is a lifelong inherited bleeding disorder. In haemophilia one of the clotting factor proteins important for blood clotting is either partly or completely missing. People with haemophilia take longer than normal for bleeding to stop. They may have bleeding into joints and muscles without having had an injury, so treatment is aimed at reducing spontaneous bleeding.

There are two types of haemophilia:

- haemophilia A is a deficiency of factor VIII (8)
- haemophilia B (also known as Christmas Disease) is a deficiency of factor IX (9).

Both types of haemophilia have the same symptoms and are inherited in the same way, though treatment is different depending on which clotting factor is missing. Specialist blood tests are needed to measure the clotting factors to show whether factor VIII or factor IX is affected and how much is missing.

von Willebrand disorder

The most common bleeding disorder is von Willebrand disorder (VWD), also known as von Willebrand disease. This is an inherited condition that can make you bleed more easily than normal. People with VWD have a low level of von Willebrand factor (VWF) in their blood, or it does not work very well. VWF works closely with factor VIII (eight) to help blood cells stick together (clot) when you bleed. If there's not enough of it or it doesn't work properly, it takes longer for bleeding to stop. There's currently no cure for VWD, but it doesn't usually cause serious problems and most people with it can live normal, active lives.

There are three types of VWD:

- Type 1 is the most common. There is a reduced amount of von Willebrand factor (VWF) in the blood.
- Type 2 is the next most common. The VWF protein is abnormal and doesn't work properly. Type 2 includes several sub-types.
- Type 3 VWD is the rarest and the most severe form. People with type 3 VWD have almost no VWF. As VWF transports factor VIII in the bloodstream, people with type 3 VWD have very low levels of factor VIII as well.

Platelet function disorders

Platelet function disorders affect a small percentage of the population. Most of these disorders are mild and many go undiagnosed. However, some platelet function disorders, such as Glanzmann thrombasthenia, are associated with more severe bleeding. Depending on the type of platelet function disorder, platelets may not stick to the walls of damaged blood vessels or form a proper surface so that other clotting factors can form a clot at the site of an injury.

All clotting factor and platelet disorders can affect girls.

We have a range of booklets available on the different types of bleeding disorders on our website.

How does blood clot normally?

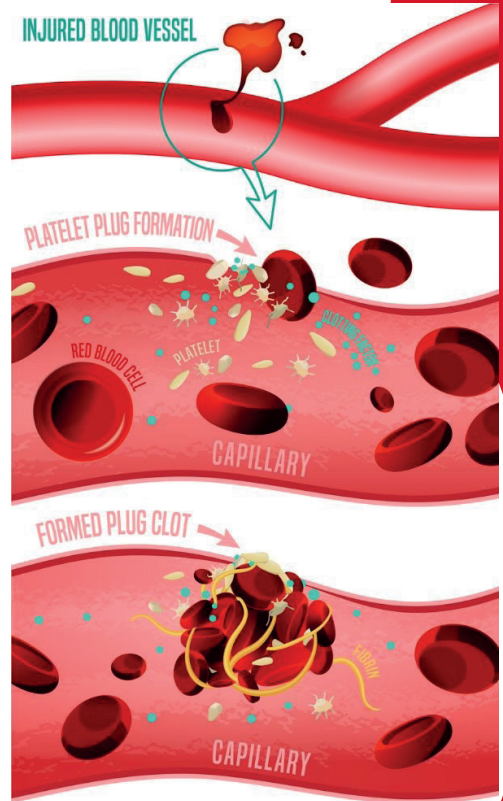
Blood is carried throughout the body in a network of blood vessels – arteries, veins and capillaries. When part of the body is injured, damage to blood vessels can cause holes in the vessel walls, where blood will leak out.

The vessels can break near the surface, as in the case of a cut, or they can break inside the body, causing a bruise or internal bleeding.

When a blood vessel is injured, the vessel walls contract to limit blood flow to the damaged area. Small blood cells called platelets are activated. They stick to the site of injury and spread along the surface of the blood vessel to stop bleeding.

The activated platelets release chemical signals that attract more platelets to the area. These clump together to form a 'platelet plug'. On the surface of the platelets, many different clotting factors work together in a series of chemical reactions. This is known as the clotting (or coagulation) cascade – it's like a chain reaction. The result is a fibrin clot, which acts as a mesh to hold the platelets together and stop the bleeding.

Normally, clotting factors circulate in the blood in an inactive form to prevent clots forming in the body when not needed. The picture above shows the stages in clot formation to make it easier to understand.



How do you get a bleeding disorder?

Bleeding disorders are generally inherited, meaning they are passed from parent to child in their genes so that you have the condition from birth.

When a baby is conceived, two sets of genes are brought together, one from each parent. Genes are small sections of DNA within the genome that code for proteins. Everything about us, from our eye colour to our height, is coded in our genes. Different sets of genes carry information for different characteristics.

Sometimes genes carry faults that adults can pass on to their children. For most bleeding disorders, you have to inherit a specific gene fault from a parent before you develop the condition. People who inherit a gene fault from one parent may be called 'carriers' but may also have the condition. Usually though, they will not have the condition but could pass the gene fault to their children. Carriers may have lower clotting factor levels than normal and may have symptoms (but also may have no symptoms at all).

Bleeding disorders are inherited by one of two patterns of inheritance:

- autosomal inheritance
- sex linked inheritance

Autosomal inheritance

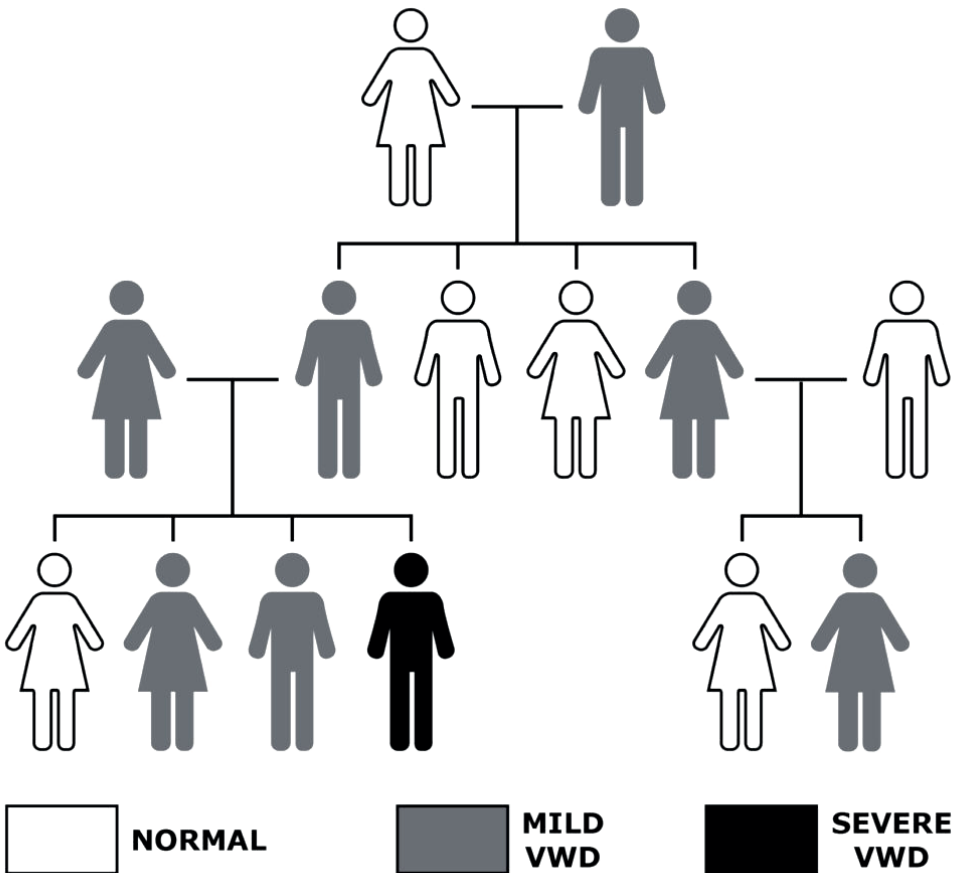
Autosomal inheritance pattern is when the faulty gene that causes a bleeding disorder is on a chromosome which does not decide the sex of the child it means the bleeding disorder is inherited in an autosomal manner. Bleeding disorders inherited in an autosomal manner affect girls as well as boys.

Most bleeding disorders have autosomal inheritance patterns, for example deficiencies of factors I, II, V, VII, X, XI, XIII, VWD and some platelet function disorders. There are two types of autosomal inheritance patterns: autosomal recessive and autosomal dominant.

Autosomal dominant means that only one defective gene, from one of the child's parents, is required for the disorder to affect the child.

Autosomal recessive means that two defective genes, one from each parent, are required for the disorder to affect the child. This means both parents must be carriers. A carrier of an autosomal disorder is someone who carries the defective gene without being affected by the disorder. The carrier of a factor deficiency may have a factor level just below, or at the lower limit of, the normal range.

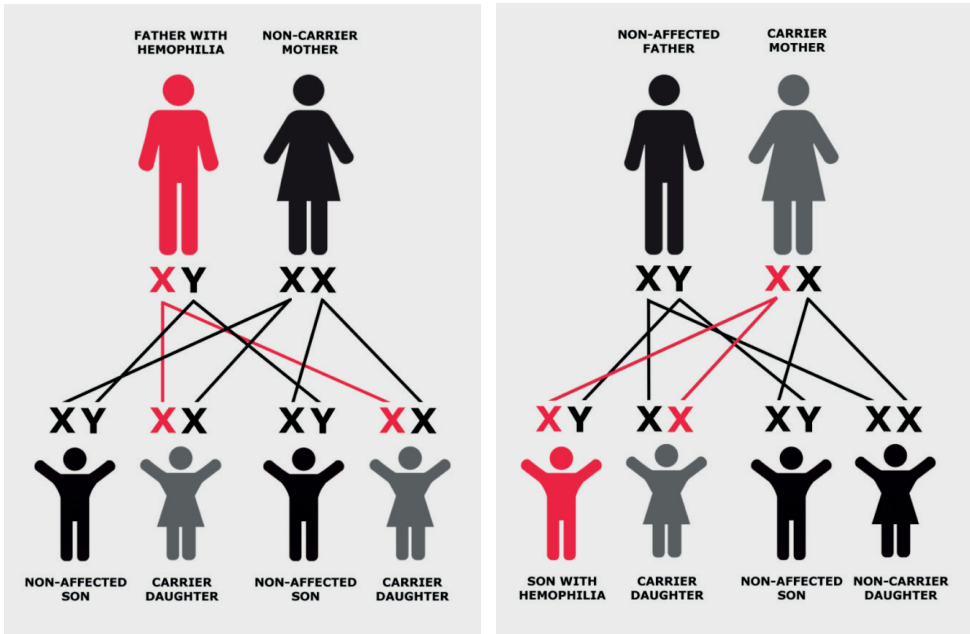
Example below is von Willebrand Disorder



(image from <https://elearning.wfh.org/wp-content/uploads/2016/06/VWD-Inheritance2-1024x946.jpg>)

Sex-linked inheritance pattern

When the defective gene that causes a factor deficiency is on the chromosome that decides the sex of a child, inheritance is said to be sex linked. The factor VIII and IX genes are on the X chromosome so haemophilia A (factor VIII deficiency) and haemophilia B (factor IX deficiency) are inherited in a sex linked manner. All girls have two X chromosomes – one from their mother and one from their father. All boys have one X chromosome from their mother and one Y chromosome from their father.



https://elearning.wfh.org/wp-content/uploads/WFH/Banners_eLearningCentres/info_how_is_hemophilia_inherited_1.png

Bleeding disorders don't run in my family, so can I have one?

Up to a third of bleeding disorders result from a 'spontaneous mutation' caused by a change in a person's genes at conception, so there won't be a history of bleeding disorders in these cases. Also, some people may not be aware of a bleeding disorder in their family if other relatives are undiagnosed or if their symptoms are different.

What symptoms do girls with a bleeding disorder have?

Symptoms will vary depending on the exact type of bleeding disorder you have but some symptoms are common to all of them. Common symptoms of bleeding disorders in girls are:

- bruising easily
- heavy periods
- pain with periods
- pain with ovulation (when an egg is released from the ovary halfway between periods)
- bleeding or oozing that lasts for a long time after surgery, dental procedures including having a tooth out, medical procedures, injuries or accidents
- bleeding from the gums, usually after trauma/injury
- frequent nosebleeds, or nosebleeds that are difficult to stop
- heavy bleeding that lasts longer than expected after childbirth
- bleeding that lasts longer than expected after cuts
- anaemia (low red blood cell count/low blood iron levels).

About half of adolescent girls with a bleeding disorder have heavy menstrual bleeding (HMB) when their periods start. Others may not until their cycles become regular and some don't have heavy periods at all.

If heavy periods are something a girl's mother or sibling has, often they are accepted as normal. But a family history of heavy periods can be an indicator of a bleeding disorder.

Rarely, girls with very low clotting factor levels (such as severe haemophilia or VWD or rare bleeding disorders) may also have:

- joint and muscle bleeds which can occur often and cause swelling and pain
- bleeding episodes that seem to happen for no obvious reason.

Because girls start having periods in puberty, symptoms of a mild or moderate bleeding disorder are often more obvious from an earlier age in girls than in boys. If you have one or more of the bleeding symptoms listed and/or a family member with a bleeding disorder (such as VWD, haemophilia, or rare bleeding disorders) you should be tested to find out if you have a bleeding disorder too. If you have heavy periods as well as other bleeding symptoms or needed iron, admission to hospital, or a blood transfusion in the past, you should also be referred for testing.

Diagnosing bleeding disorders

Depending on the type of bleeding disorder, you may know you have one from birth, or you may be diagnosed as a child or young adult.

Some bleeding disorders can have mild symptoms or no symptoms at all. So you may not find out until you are an adult, perhaps after having abnormally heavy bleeding when having a tooth out or after having an operation.

To make a diagnosis you will need blood tests. These tests can't be done in GP surgeries – they need special laboratories. The blood tests you have may include:

- full blood count (FBC) to check the haemoglobin and platelet count
- clotting screen or coagulation screen
- clotting factor levels
- von Willebrand factor investigations
- platelet function studies.



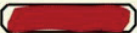
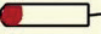

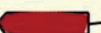
The blood sample will be taken from the front of your arm near the bend at your elbow. If it's difficult to find a vein there, the back of your hand may be used instead.

Diagnosing bleeding disorders is complex, particularly if you have no known family history. You may need to have blood tests repeated as the levels in the blood can be affected by anxiety, your menstrual cycle and any medicines you may be taking. Although this can be frustrating it's nothing to worry about and you can be sure that full investigations are being done to find out what is causing your bleeding. Once you have a diagnosis, other family members can be offered testing. Once you have a diagnosis, other family members can be offered testing. It's easier for them to get a diagnosis as the bleeding disorders team know what condition they are looking for. Your hospital will contact you as soon as they have the results back from the blood tests.

What to do if you think you may have an inherited bleeding disorder

The first thing to do is to see your GP and ask to be referred to a haematologist (a doctor who specialises in blood diseases). Many GPs may not be familiar with bleeding disorders so it's important to keep a record of your periods. This includes

when you bleed, how long your periods last, how much you are bleeding each day, pain during periods, when you spot bleed, when you get pain etc. like the example here:

Date: Week 5/11		5/18		Days of bleeding								Number of used sanitary towel
Sanitary towel				1	2	3	4	5	6	7	8	
Intensity of bleeding per sanitary towel Factor:	x 1		//	/	/	/	//	/				
	x 5			###	///	//						
	x 20			//	//							
and/or Tampons												Number of used Tampons
Intensity of bleeding per tampon Factor:	x 1			/			/					
	x 5			//	///	//						
	x 15			###/	///							
Daily points:				2	137	101	21	3	1			= 265

Or you can use a menstrual bleeding app on your mobile phone to help track bleeding such as Flo Period and Ovulation, Period tracker or Eve by Glow.

You might find it intimidating to tell a medical professional that you think you have a bleeding disorder, but don't be afraid to speak out. Take a parent or older sibling with you for support. It is important to get a proper diagnosis so that you can receive sufficient treatment, care and support.

Genetic counselling and testing

Genetic counselling is an essential but complex part of proper care for individuals and families with a bleeding disorder. Genetic counselling should consider your experiences and perceptions, as well as the social, cultural, and religious factors and contexts that may influence your decisions and the options you have.

Genetic testing can work out the exact genetic mutation causing a certain condition. For bleeding disorders, genetic tests may answer the following questions:

- If you know you have a bleeding disorder, what is the genetic change that has caused your condition?
- Are you a carrier of a bleeding disorder?

Depending on the type of disorder and the reason for testing, the genetic test results can take weeks or months. Your haemophilia centre may be able to give you an estimate of the time it will take.

Carrier testing

Unlike factor testing, it's less clear when genetic tests should be done. Views differ on the advantages and disadvantages of testing at an early age and this is something families should discuss with their bleeding disorders team.

However, it's important for young women and girls to know if they are carriers well before they have any thoughts of having children. Knowing this will make it easier for planning pregnancies. There are two tests that can be done to find out if you are a carrier. Firstly, there is a genetic test to determine carrier status. Secondly, there is a factor level test to measure factor levels. This can help determine whether there is a greater than normal risk of bleeding.

It is important to know the factor level in a potential carrier due to the increased risk of bleeding that you may experience with low factor levels, i.e. bleeding after tooth extraction, trauma or surgery. Also when you have your period there is a tendency for excessive bleeding and if you have very low factor levels you may require medical treatment. Carriers may also need medical intervention for nosebleeds. If levels are low, precautions can be taken to prevent many of these complications.

As a carrier, you may experience a range of emotional and psychosocial impacts related to having your own family or the consequences of passing on a genetic disorder. Your experience of being a carrier may change with different life stages, and you may need genetic and/or psychosocial counselling and support more than once in your life.

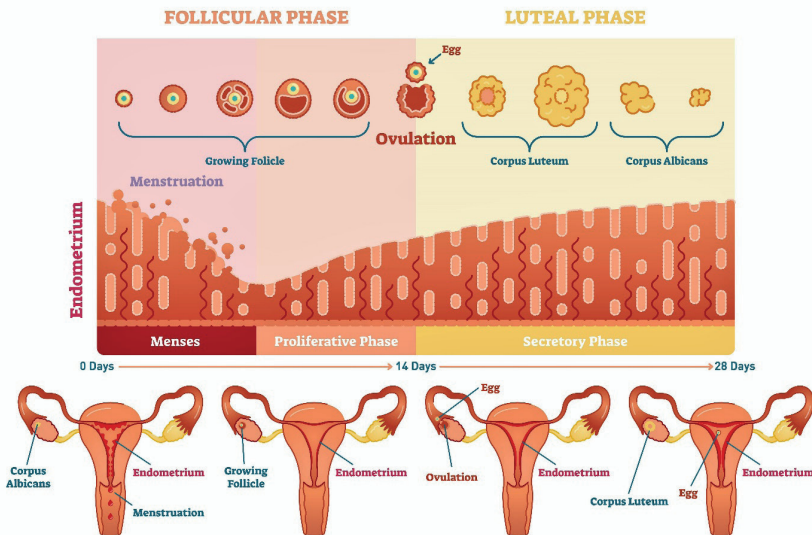
Please remember that if you are diagnosed with a bleeding disorder, it might at first seem overwhelming. But we want to reassure you that with ever better treatments and care available, people with bleeding disorders usually manage their conditions very well and can get on with living and enjoying their lives.

Section 2 – About periods

Our bodies change as we grow older. It is all part of growing up. Your body may do things you're not familiar with. The best thing you can do is to learn what to expect. Puberty is the physical and emotional changes your body goes through on its way to adulthood, and for girls, it usually starts between ages 8 and 13. The average age for girls to begin puberty is 11 years.

One of the biggest changes for girls during puberty is starting their periods. You might hear this called 'menstruation'. A menstrual cycle is on average about 28 days, but it can be slightly less or more than this. The cycle stops while a woman is pregnant.

These are the main features and changes during the menstrual cycle.



The start of the cycle, day 1, is when bleeding from the vagina begins. This is caused by the loss of the lining of the uterus (womb) with a bit of blood. This is your period. By the end of about day 5, the loss of blood stops. The uterus lining begins to regrow, and an egg cell starts to mature in one of the ovaries.

At about day 14, the mature egg cell is released from the ovary. This is called ovulation. The egg cell travels through a tube called the fallopian tube toward the uterus. If the egg cell does

not meet with a sperm cell in the oviduct, the uterus lining begins to break down, and the cycle repeats.

About menstrual cycles

Ovulation pain

Some girls get a one-sided pain in their lower abdomen (tummy) when they ovulate. It happens about 14 days before your period when the egg is released from the ovary, so is sometimes known as 'mid-cycle pain'. The pain can be a dull cramp or a sharp and sudden pain, and it can be on either the left- or right-hand side of your tummy, depending on which ovary is releasing the egg, but sometime the pain is generalised all over your tummy. It can last just a few minutes or continue for a day or two. Painful ovulation can usually be eased by simple remedies like soaking in a hot bath or taking an over-the-counter painkiller, like paracetamol. You may notice slight vaginal bleeding when it happens.

Painful ovulation is fairly common and usually harmless. See your GP if the pain is severe or you're worried, as it can sometimes be a symptom of a medical problem. It's a good idea to keep a diary before your visit. Let the doctor know exactly when during your menstrual cycle the pain comes on and how long it lasts.

Ovulation bleeding

Ovulation bleeding is light spotting that occurs when your ovary releases an egg. It can happen during the middle of your cycle. For some girls, changes in hormone levels can trigger light vaginal spotting during ovulation. Ovulation doesn't go on for very long, and neither does the bleeding – a day or two at most. Bleeding tends to be very light and you may only need to use a panty liner. The blood is usually pale pink; that's because the blood is mixed with cervical fluid, which increases during ovulation.

Ovulation can also be associated with slight bleeding from the ovary as it releases the egg and can cause abdominal pain called mid-cycle pain (discussed above). In girls with severe bleeding disorders this bleeding can be severe and need immediate medical attention. If you have a severe pain or bleeding or feel faint, you should contact your haemophilia centre.

Periods

Girls with bleeding disorders might have more symptoms during their monthly periods. Heavy periods that last longer than normal are the most common symptom for girls with bleeding disorders. This is called heavy menstrual bleeding (HMB), which doctors used to call menorrhagia.

HMB is defined as excessive menstrual blood loss which interferes with a woman's physical, social, emotional and/or material quality of life. It can occur alone or in combination with other symptoms.



If your period lasts for more than seven days, you are soaking through sanitary protection (pads or tampons) every two hours or less, or you have clots that are bigger the size of a 10p coin, then you should contact your GP or haemophilia centre. Every girl is different, and what is considered 'normal' bleeding during a period for one girl may be 'excessive' for another. There is no easy way to measure the exact amount of blood you lose during your periods, so it's important to keep a record of number of days you bleed, how many pads/tampons you use and how bloodstained they are, or the amount of blood in the menstrual cup if you are using one. There are also apps you can use on your mobile phone for logging your symptoms.

Girls can also have heavy, irregular periods when they first start having them. This may improve as things settle down. If you continue to have heavy menstrual bleeding, keep a diary of your periods (track your blood loss during your period), as described in page 13.

What is a normal period?

Gynaecologists often define a normal period as:

- bleeding for around 5 to 6 days up to about 7 days
- losing about 30 to 40mls of blood per month (around 2 to 3 tablespoons).



- bleeding every 25 to 35 days
- average cycle is 28 days.

What is an abnormal period?

- losing 5 to 6 tablespoons (about 80ml or more) of blood
- bleeding lasting longer than 7 days
- passing lots of large clots and flooding
- erratic periods
- interferes with your physical, social, emotional and/or quality of life.

You may experience heavy periods if you have been diagnosed with the following:

- VWD
- factor deficiencies – haemophilia A and haemophilia B
- other rare bleeding disorders (factor II, V, VII, X, XI, XIII)
- fibrinogen or prothrombin deficiency
- thrombocytopenia (including congenital or acquired) including ITP and TTP
- other rare bleeding disorders e.g. Glanzmann thrombasthenia, Bernard-Soulier syndrome, platelet dysfunction.

Living with heavy menstrual bleeding shouldn't be dismissed as a minor issue. Heavy periods can affect your health, sense of wellbeing, and daily life. If you bleed a lot during your periods and you know other girls in your family do too, you might not realise the amount of blood lost during your monthly period is more than is normal in most girls. But there are treatments that can help – find out more on page 22.

Anaemia

Heavier and longer monthly periods can lead to anaemia (low levels of red blood cells), causing weakness and tiredness. Because blood contains iron, girls with bleeding disorders can have low levels of iron in their blood. Girls should have their blood tested regularly for anaemia. Iron supplements may be prescribed for this.

Diet is important to help with anaemia. Try to eat plenty of foods rich in iron (e.g. red meat and green vegetables) or take iron supplements to help keep your iron levels up. Also speak to your GP/haemophilia team if this persists.

What sanitary products should I use during my periods?

These days there are a lot of options. You can try different options and combinations until you find what works best for you.

Pads: A pad (sometimes called a sanitary towel) is an oblong-shaped, absorbent material that you stick to the inside of your underwear to soak up period blood. Some pads have extra material folds on the sides, called wings that you fold over the sides of your underwear to help keep the pad in place and prevent leaks. Pads come in different lengths and sizes to provide more or less protection as needed.

Panty liners: Panty liners are similar in shape to pads, but they don't absorb as much blood because they are thinner. Some girls use them as protection on lighter menstrual flow days or as back-up protection with tampons.

Tampons: Tampons are made from absorbent material in a tube shape that can be pushed up into the vagina to soak up the blood. You also throw them in the bin after use. Each tampon should not be used for more than eight hours. You may need to change them more often if they are saturated. They come in different sizes. They can have plastic or cardboard applicators to help insert them in the vagina. A tampon won't hurt if inserted correctly, and you won't really feel it. Some girls find these tricky to put in at first, but others prefer them, especially when playing sports or swimming.

Menstrual cups: A menstrual or moon cup is a small, flexible cup inserted into the vagina to catch and collect blood. It doesn't absorb the blood like a pad or tampon; instead, it forms a seal so no blood will leak out. A menstrual cup is safe to use from your first period although it may feel a little uncomfortable at first. You can wear a menstrual cup for 6 to 12 hours, depending on whether you have a heavy flow. This means you can use a cup overnight. Reusable menstrual cups should be washed and wiped clean before being reinserted into your vagina.

Period underwear: Period underwear is made of special absorbent material that you can wear like normal underwear. Depending on how heavy your flow is on a given day, it can last most or all day or can be a back-up for a tampon. You may need to have more than one pair to make it through your periods, but they are reusable month after month.

Managing periods in the classroom

Any girl just starting her periods may have heavy bleeding. Heavy menstrual bleeding (HMB) is also common among girls with bleeding disorders and varies in what is considered 'normal' blood loss or pain during periods. This can have a big impact on your everyday life such as school and your social life.



Girls with heavier blood loss may need to use the toilet or medical room regularly, which may mean you need to leave lessons more often. You should be given a toilet pass, and teachers made aware that you may frequently leave the classroom during your period. Understanding and privacy are essential, especially as leakage may show through clothing. You should keep a small bag at school with spare clothing if you need to change and have a good supply of tampons/pads.

If you feel tired or look pale during your period, you shouldn't take part in physical activities at school at that time. You can be positively involved in PE lessons: ask your teacher if you could umpire sports games or assist with timing of games, cross country running etc. Light exercise may help relieve the tension in the muscles and ease the pain caused by cramps, so there may be alternative activities you could do. It's best not to swim if you are concerned about leaking blood even when wearing a tampon.

No two girls experience the same period, so it's important for your school to show understanding and to remember that as a girl with a bleeding disorder, you may be embarrassed to talk about yours. It might be helpful to have a nominated person or first aider who could help ensure your needs are being met and that staff respond appropriately.

Top Tips for managing HMB

by Jessica Page, THS Youth Ambassador



Growing up, I struggled with bad bruises and heavy bleeding. My dad has severe haemophilia A, but I was always told there was nothing wrong with me, as 'girls couldn't have haemophilia'. It wasn't until I was 18 that I was finally tested and confirmed that my factor levels were low enough to be diagnosed with mild haemophilia. After a while, we decided that my symptoms didn't seem to match up that of a mild haemophiliac, so at the age of 25 I was also diagnosed with VWD! I enjoy being a youth ambassador because I want to support others in our community and raise awareness of bleeding disorders in girls.

I thought of 'top tips' for managing my period that might be helpful for others. I hope you find them useful.

- Avoid wearing white or lighter coloured clothes around the time of your period.
- Have 'period pants' – underwear you only wear when you're on your period. This prevents your newer, nicer underwear from being stained and ruined.
- Experiment with different kinds of protection (in different combinations) to find what works for you: tampons, menstrual/moon cups, period underwear, sanitary pads/towels etc.
- Cotton or organic sanitary towels can be kinder to your skin than alternatives, which may be more appropriate for longer periods, though they can be more expensive.
- You can buy bedsheets and mats that will help protect your bedsheets and provide another layer between you and the mattress for added overnight protection.
- Have an 'emergency' pack at work or school (or in your car) that contains spare underwear, sanitary protection and maybe even spare clothes.
- Carry a disposable or zip-lock/sandwich bag with you so you can dispose of your used sanitary protection if there aren't any bins available
- Most people find tampons more comfortable for sports activities. Change it before and after your physical activity for the best protection. If you prefer wearing pads, tighter underwear can stop it from moving around and becoming uncomfortable as you move.
- Wearing 'boy short' pants or shorts over your knickers can help you feel more comfortable and give you another layer of protection.
- Carry a period bag around with you inside your handbag or rucksack. This bag should include sanitary provision, painkillers, spare knickers and anything else that you may need. Handy if you're caught out!
- As well as painkillers, heat packs and hot water bottles can help with cramping. You can also now buy heat patches for a more discrete alternative.
- Eating extra snacks while you're on your period can help to manage tiredness.
- Make sure someone senior at school or work knows about your bleeding disorder so you have the confidence to know that you may need to leave meetings/lessons/work during the day to change your sanitary protection more often.
- Go to your GP or haemophilia centre if you are struggling with your periods and they are starting to impact your everyday life.

Section 3 – What are my treatment options?

Bleeding such as bruising, minor cuts and some nosebleeds can often be controlled using simple first aid measures such as RICE (Rest, Ice, Compression and Elevation). If bleeding continues in cases like heavy or long-lasting nosebleeds, some dental procedures, during or after surgery or after injury, other treatment options may be needed. Bruising may also need treatment, especially if it is extensive or painful, or there are raised haematomas (where blood collects in the tissues under the skin).

Periods will occur every month, unfortunately there is nothing we can do to stop this! However, for girls who experience heavy menstrual bleeding there are also hormonal treatment options that will help.

Non-hormonal

Tranexamic acid (Cyklokapron)

Tranexamic acid is an anti-fibrinolytic agent. This means that it slows the breakdown of blood clots. It is often used to prevent or treat bleeding from mucous membranes such as the inside of the mouth, nose, gut or womb. It is usually taken as tablets, three or four times a day, but may also be given as an intravenous infusion (drip into a vein). You will be able to get a repeat prescription for tranexamic acid from your GP/haemophilia centre.

A liquid form is also available for children, though you will usually need to get this from your haemophilia centre.

Tranexamic acid may be given before some dental treatments, for nosebleeds or prolonged or heavy periods. It may be used alone or in combination with DDAVP[®] and/or factor replacement therapy. Tranexamic acid does not replace or increase clotting factor levels. This means it cannot be used instead of DDAVP[®] or factor concentrate, but for some minor procedures it may be the only treatment needed.

It can sometimes have side effects including:

- nausea (feeling sick)
- dizziness
- diarrhoea (watery or loose poo)
- stomach pain.

For heavy periods, you should take tranexamic acid as soon as your period starts. This reduces the breakdown of a pre-formed blood clot, which reduces bleeding. You may need to take it three times a day for around 3 to 5 days (or longer if needed) to reduce blood loss.

Desmopressin (DDAVP®)

DDAVP® is a synthetic drug that can be suitable for some people with milder forms of haemophilia A or VWD. It releases factor VIII stored in the lining of blood vessels, increasing the amount of factor VIII circulating in the blood. This increase can be enough to control minor bleeding episodes and to prevent bleeding from minor operations including dentistry. It is given as a subcutaneous injection (under the skin like a vaccination) or as a nasal spray. It can't work for severe haemophilia as there are no stores of factor VIII. For some girls, desmopressin is a helpful treatment for heavy periods.

Clotting factor replacement

For girls with severe factor deficiencies that don't respond to other treatments, regular prophylaxis with clotting factor replacement may be needed to control heavy menstrual bleeding. These may be needed when the risk of severe bleeding is high, such as during childbirth, before major surgery or there has been a head injury.

Iron supplements

You may be advised to take iron supplements to boost iron levels in the blood. If your iron levels don't improve, you may need an intravenous infusion (drip into a vein) of iron. These are given in a hospital clinic as there is a risk of an allergic reaction.

Hormonal

Combined hormone contraception (CHC)

Hormones in CHCs work by preventing the ovaries from releasing an egg. They also thin the lining of the womb which often makes periods lighter and less painful. These can come as pills, a patch or a vaginal ring. CHCs prescribed as an effective treatment for heavy periods. You may be advised to use this continuously for a few months so that you don't get a period.

Progestogen-only pill or cyclical progestogens

Some progestogens can be used to make periods lighter and less painful such as cyclical progestogens (e.g. norethisterone or medroxyprogesterone acetate), a desogestrel

progestogen-only pill and Depo-subQ. The progestogen-only pill needs to be taken every day to work. You need to take one pill every day within either 3 or 12 hours of the same time each day, depending on which type you are taking.

If you're a young teenager, you may be nervous that your parents may not want you to use hormonal contraceptives. A good discussion with your haemophilia team can provide you and your parents with accurate information and help you all work through any concerns.

Intrauterine system (IUS)

An IUS is a small T-shaped plastic device that releases the hormone progestogen directly into the womb, which thins the womb lining to reduce heavy periods. Brand names include Mirena and Levosert. It can give you some erratic bleeding at first but this device lasts for up to five years, depending on the brand.

Advantages

- The IUS is inserted by a doctor or nurse in a clinic and does not need an operation.
- If you are having sex, the IUS is an effective method of birth control, though you can still get pregnant after it is removed.
- You don't have to remember to take pills on a regular basis.

Disadvantages

- Some women experience discomfort and cramping when the IUS is put in.
- Periods can be irregular for the first 3 to 6 months after the IUS is put in.

If one treatment doesn't suit you, there are others you can try.

Section 4 – Living with a bleeding disorder

What does having a bleeding disorder mean to you?

By the time you reach your teens, you may already have a lot of experience talking about your condition to friends and schoolmates. Some girls are diagnosed with bleeding disorders in their teens or even later in life. Later diagnosis may add extra anxiety to what can be a stressful time for anyone.

At first, you may find talking about your bleeding disorder difficult. The hardest part is probably bringing the subject up. When you tell people, and how much you tell them, it is entirely up to you. They may find it challenging to take it all in at first but remember, you don't have to say everything in one go.

How you tell people can make a difference. Most people will want to know more and are likely to have questions, so you will need time to have a proper chat. Try to be optimistic about your condition. If you sound confident when you talk about managing it, they will be confident too.

People who care about you are not likely to react badly – if they do, it may be because it can sound a bit frightening when they first hear about it. Many people will never have heard of bleeding disorders. Once they've had a chance to think about it and ask you more about what it's like, most people will be positive and realise that it's just another part of your life.

Exercise

Exercise and sport have many benefits for health and can improve self-esteem, learning and concentration. If you have a bleeding disorder there are some particular benefits: strong muscles, good balance, and posture can help protect joints from

bleeding. Maintaining a healthy weight helps to reduce stress on joints that have already been damaged by bleeding. If you have heavy periods and are nervous about wearing gym kit for fear of bleeding through a tampon or pad, ask if you can wear something else – and read the top tips on page 21.

Your choice of activity or sport will be individual, and your haemophilia team will discuss the risks and benefits of different sports taking account of your diagnosis. In general, sports involving a lot of physical contact and those where head and neck injuries occur carry the highest risk of injury and risk of bleeding. Any treatment you have can be tailored around days when you're most active so that you have maximum protection from bleeding at these times, especially if you get regular treatment that you can give yourself.

Tips about exercise

- Be ready for accidents, just in case: always have a first aid kit on hand (this might include emergency treatment product); wear a medical alert bracelet; and tell someone with you what to do if you hurt yourself.
- Treat injuries, bruising or bleeding promptly and give yourself time to recover fully.
- Common sense is very important – set your own achievable limits and make sure that your parents are happy with this.

Dental care

It's essential to look after your teeth and pay attention to mouth hygiene. This will help to lower your risk of gum disease and tooth decay and minimise the need for dental treatment later in life. You should:

- brush your teeth at least twice a day
- floss your teeth daily
- use toothpaste that contains fluoride
- have regular dental check-ups.

Your bleeding disorder may mean that your gums bleed a little more easily and for a little longer until the gums heal. It is important during this time to continue tooth brushing with a soft brush to remove the plaque and food debris which causes the gum disease.

If you have a mild bleeding disorder, it's okay for you to attend your dentist for routine care such as check-ups and fillings. More invasive treatment such as having a tooth out can cause a lot of bleeding. Extractions should always be co-ordinated by your haemophilia centre, this ensures that the procedure is carried out safely and prevents excessive bleeding. Depending on the severity of your bleeding disorder your haemophilia team may choose to give you factor replacement therapy and/or tranexamic acid. In addition, the dentist may stitch the wound and use a special surgical packing to stop any bleeding.

Frequently asked questions

How do I educate my daughter about her bleeding disorder?

Involving your daughter in learning about her bleeding disorder is an important part of her growth and development. As she grows up, you can teach her about her bleeding disorder, the treatment she uses, medicines she should avoid, when to seek treatment, genetic inheritance and how to speak out and get help to achieve what she needs. The type of information and how you teach it will depend on her stage of development – your haemophilia centre can help you with this.

This will be a gradual learning process throughout her childhood, which gives her an opportunity to understand and adapt to her bleeding disorder and take responsibility for managing it. When she reaches the age where she manages her own health, she will then have the information and the confidence to make independent decisions about her health and life choices.

What painkillers can I use?

Some medicines can affect blood clotting and so may not be suitable for someone with a bleeding disorder. These include aspirin and ibuprofen, which should never be taken unless advised by a haemophilia specialist. Any new medicine, including ones that can be bought over the counter and herbal preparations, needs to be considered for any increased risk of bleeding.

Some medicines can affect blood clotting and so may not be suitable, such as non-steroidal anti-inflammatory drugs (NSAIDs) like ibuprofen and aspirin. Paracetamol is suitable for treating pain and fever. Paracetamol is a suitable painkiller if you have a bleeding disorder.

Before you start taking anything prescribed by your doctor or other health practitioner check with them whether it is safe for someone with a bleeding disorder. Some medicines, vitamins and supplements can interfere with blood clotting and healing or can irritate your nasal passages or stomach lining. This includes some herbal and homeopathic medicines.

If you are uncertain about whether any medicine may affect blood clotting, it's important to check with your haemophilia team before taking it.

Do I need a bleeding disorder card?

You should always carry a 'bleeding disorder card', which is provided by your haemophilia centre. This will detail the bleeding disorder you have, treatment needed and the haemophilia centre's contact details. Haemophilia centres should be able to provide you with as many cards as you need, for example, so that one can always be left at school or college (with medicines if appropriate).

Can I have tattoos and piercings?

If you are under 18 you must have permission from your parents to get a tattoo or piercing.

If you are thinking of getting a tattoo or piercing, the type of bleeding disorder and the severity you have may mean the need to treat before and after, particularly if you have a severe bleeding disorder. Speak to your haemophilia nurse to ensure that you take the necessary precautions beforehand.

You may be discouraged from getting tattoos or piercings because of the associated medical risks, such as infection. If you decide to go ahead, it's crucial to find a reputable tattoo and piercing shop to avoid the risk of getting an infection. You should tell the person doing your piercing or tattoo that you have a bleeding disorder so they are aware of any potential complications.

There are other issues to be aware of too. Did you know getting a tongue, lip or cheek piercing can result in gum problems and tongue piercing can damage the enamel on your teeth? And some parts of the body take longer to heal than others – for example it can take up to a year for a belly button to heal properly following piercing. This means you are susceptible to infections for up to a year after the piercing is removed.

What's the best way to remove unwanted hair?

Hair growth on the body is an unwanted side effect of puberty. Although boys can be as hairy as they wish, this is not a luxury girls have. Therefore, girls have a constant battle about hair removal. There are many different hair removal methods, but each carries hazards for girls with bleeding disorders.

Shaving can easily cause anyone to cut themselves. These small cuts can be more inconvenient and take longer to stop for a person with a bleeding disorder. Ways to avoid these mishaps are to avoid using cheap or old razors - if using a disposable razor, replace it after each use. If using a non-disposable razor, ensure that the razor head is clean and rust-free. Take your time while shaving to try and avoid cutting your skin. Take particular care when shaving around the bikini line as this area is more sensitive. If you nick yourself when shaving, you should allow yourself plenty of time to stop the bleeding before leaving the house. Whether home waxing or having a professional carry out the treatment, waxing can be painful. If getting waxing done in a salon, inform the beauty therapist of your condition. It is important to carry out a patch test several hours before waxing to ensure the wax you use is suitable for your skin.

Waxing may cause small blood blisters as the skin is stretched and the hair forcibly removed from the follicle. Never wax for the first time before an important event in case bruising or blistering occurs.

Tweezing involves pulling hair out of the follicle, which can cause blistering. When tweezing eyebrows, it is important to take care as bruising or blistering will be more noticeable on the face.

Alternative methods of hair removal include hair removal creams and laser hair removal. Hair removal creams are safer than shaving, but a patch test is required 24 hours before use. Laser hair removal is significantly more expensive than other hair removal methods but results last longer. It is important to notify the beauty therapist of your condition before laser hair removal, as blistering of the skin may occur.

You are not alone

Many girls often comment that it's helpful to talk to others in a similar situation and know that they are not alone, even though bleeding disorders may only affect them at certain times in their life.

Talking Red is Haemophilia Society's campaign that urges everyone – boys and girls – to talk about girls bleeding.

We started **Talking Red** to empower girls, share knowledge and spread the word that help and support are out there.

Girls' bleeding disorders can have a significant impact mentally, physically and financially. Yet many people still don't know that girls can be affected by bleeding disorders.

Thousands of girls struggle with common symptoms – such as heavy periods and frequent bruising – unaware that they can ask to be tested for a bleeding disorder.

There is no cure for a genetic bleeding disorder, but there is effective treatment available. That's why it's so important to get the right diagnosis.

Living with a bleeding disorder can be challenging to manage and affects relationships, education and work. It can be lonely living with a condition that some people find hard to talk about.

No girl should go through this alone. **Talking Red** is here to bring people together and share knowledge and experience to empower girls to get their treatment and care.

Why don't you join our community and help get everyone **Talking Red?**

European principles of care for women and girls with bleeding disorders

The 10 principles of care listed below are a benchmark for diagnosis and management of women and girls with bleeding disorders. They aim to improve awareness of their unique challenges and positively impact their health, wellbeing and quality of life.



EUROPEAN PRINCIPLES OF CARE FOR WOMEN AND GIRLS WITH INHERITED BLEEDING DISORDERS

Women and Girls with Bleeding Disorders (WGBD) face unique challenges. In order to optimise diagnosis, care and management of WGBD, the EAHAD Women and Bleeding Disorders Working Group have developed the following Principles of Care (PoC):



<p>Equitable access and quality of care for all individuals with bleeding disorders, irrespective of gender.</p>	<p>Timely and accurate diagnosis of bleeding disorders in women and girls</p>	<p>Awareness of the additional challenges faced by WGBD throughout life</p>	<p>Comprehensive care of bleeding disorders requires a family centred approach which includes WGBD</p>	<p>Inclusion of a dedicated obstetrician and gynaecologist in the multidisciplinary team</p>
<p>Education of WGBD and their families regarding the menstrual cycle and management</p>	<p>Early recognition and optimal management of heavy menstrual bleeding</p>	<p>Provision of preconception counselling and access to prenatal diagnostics</p>	<p>Provision of a patient centred comprehensive management plan throughout pregnancy and the post partum period</p>	<p>Involvement of WGBD in registries, clinical research and innovation</p>



THESE PRINCIPLES OF CARE SERVE AS A FRAMEWORK TO GUIDE HAEMOPHILIA TREATMENT CENTRES WITH THE AIM OF POSITIVELY IMPACTING ON THE HEALTH, WELLBEING AND QUALITY OF LIFE OF WGBD



You can read more about the 10 principles of care on our website at:
<https://haemophilia.org.uk/bleeding-disorders/women-with-bleeding-disorders/european-principles-of-care-for-women-and-girls-with-bleeding-disorders/>

Glossary

Acne

Bumps on your skin (especially your face, neck, back and upper arms) caused by extra oil your skin makes. Acne is also called 'pimples' or 'zits'.

Adenomyosis

The tissue that normally lines the womb starts to grow within the muscular womb wall, making your periods particularly painful.

Bernard Soulier syndrome

A disorder caused by a missing or non-working protein on the surface of a platelet called the glycoprotein Ib/IX/V.

Carrier

A person who has the gene for a condition but doesn't always have the symptoms for the condition.

Chromosome

The chromosomes are found in the nucleus of each cell. Each cell with a nucleus contains chromosomes, which are made from DNA. Human body cells each contain 23 pairs of chromosomes, half of which are from each parent.

Factor I (1) deficiency

A bleeding disorder in which the body doesn't have enough of the blood protein fibrinogen or none at all or the protein does not work correctly.

Factor II (2) deficiency

A bleeding disorder in which the body doesn't have enough of the blood protein prothrombin or none at all or the protein does not work correctly.

Factor VII (7) deficiency

A bleeding disorder in which the body doesn't have enough of factor VII or none at all or factor VII does not work correctly.

Factor XI (11) deficiency

A bleeding disorder in which the body doesn't have enough of factor XI or none at all or factor XI does not work correctly.

Factor XIII (13) deficiency

A bleeding disorder in which the body does not have enough of factor XIII or none at all or factor XIII does not work correctly.

Fallopian tube

The part inside your body that carries the eggs from your ovaries to the uterus (womb).

Glanzmann thrombasthenia

A condition caused by a missing or non-working protein on the surface of a platelet called the glycoprotein IIb/IIIa.

Gynaecologist

A doctor who specialises in the female reproductive system, including menstruation (periods), reproductive medicine, contraception (birth control) and menopause.

Haematologist

A doctor who specialises in researching, diagnosing, treating and preventing blood disorders.

Haemophilia A (factor VIII deficiency)

A genetic disorder caused by missing or non-working factor VIII, a protein needed for the blood to clot.

Haemophilia B (factor IX deficiency)

A genetic disorder caused by missing or non-working protein called factor IX, making it difficult for the blood to clot. Haemophilia B is also known as factor IX (FIX) deficiency.

Heavy menstrual bleeding (HMB)

Menstrual periods with abnormally heavy bleeding that limits daily activities and places girls at increased risk of health problems if left untreated.

Intrauterine system (IUS)

A small, T-shaped plastic device that's put into your womb (uterus) by a doctor or nurse to stop you getting pregnant.

Menstrual cup

A small, flexible cup that is inserted into the vagina to collect period blood. Also known as a moon cup.

Menstrual cycle

The number of days from the start of one period to the start of the next period. Younger girls may experience cycles every 28 to 45 days. On average, the menstrual cycle is 28 days but can range from 21 to 35 days.

Menstruation

The medical term for a period.

Ovaries

The part inside your body that makes eggs.

Period

The time of the month when the womb (uterus) lining sheds and blood comes out of your vagina. Normal periods usually last 2 to 7 days.

Puberty

The physical and emotional changes your body goes through on its way to adulthood, usually starting between ages 8 and 13 for girls.

Progestogens

Synthetic forms of progesterone.

Prophylaxis

Treatment is given regularly to prevent bleeding

Thrombocytopenia

You have fewer platelets than would be optimal.

Vagina

A muscular tube connecting the womb (uterus) to the outside of the body.

Vaginal discharge

Thick, clear or white fluid that comes out of the vagina to help maintain a healthy vagina.

von Willebrand disease (VWD)

A disorder that is caused by a lack of or a problem with the von Willebrand factor (VWF) in the blood.

von Willebrand factor (VWF)

A clotting protein that helps platelets stick together to stop bleeding; factor VIII is attached to VWF.



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The Haemophilia Society makes every effort to make sure that its services provide up-to-date, unbiased and accurate information about bleeding disorders.

We hope that this information will add to the medical advice you have received and help you to take part in decisions related to your treatment and care. Please do continue to talk to your doctor or specialist nurse if you are worried about any medical issues.

Give us your feedback

If you have any comments about this booklet or any of our other information, please write to the Head of Policy and Programmes at the address below.

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