

Teenage Girls with Bleeding Disorders



Introduction

It is often mistakenly assumed that haemophilia and other bleeding disorders occur almost exclusively in males. It is a fact that classic haemophilia A and haemophilia B in their severe form occur primarily in men, however women who are carriers of the factor VIII or factor IX gene can often have low levels and can have symptoms due to their bleeding disorder. In addition, in Ireland there are more females than males diagnosed with von Willebrand Disease, with some of the rarer bleeding disorders and with bleeding disorders where the type is unknown.

For teenage girls there are many specific and relevant issues which may be of concern and for which they require information. A bleeding disorder can have an impact on menstruation and on preparation for childbirth. They need to get an understanding of treatment options. There are issues to be considered in relation to relationships, tattoos, piercings and personal care.

We hope this information booklet will be informative, educational and useful in a practical sense to all teenage girls with bleeding disorders in Ireland.

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Common Bleeding Disorders

Haemophilia:

Haemophilia A (factor VIII deficiency) and haemophilia B (factor IX deficiency) mostly affect males, but it is possible for women to have haemophilia. There are three levels of severity in haemophilia, mild (factor levels of 5% or more), moderate (factor levels between 1% and 5%) and severe (factor levels less than 1%). Severe haemophilia in women is extremely rare.

Haemophilia is a sex-linked genetic disorder, which means women are generally carriers of haemophilia. Two thirds of female carriers of haemophilia have normal levels and do not have any bleeding problems. However, a third of female carriers have low levels, less than 40%, and may have some bleeding symptoms for example heavy periods or bleeding after surgery.

Von Willebrand Disease:

Von Willebrand disease (vWD) is the most common bleeding disorder that women have. It affects about 1 in 100 people. There are a number of different types and all are caused by a problem with the von Willebrand Factor (vWF). This is a protein in blood which is necessary for blood coagulation, or clotting. It helps platelets stick to the wall of a damaged blood vessel. The vWF has a second function. It carries factor VIII (another important protein in blood clotting), through the bloodstream. When there is not enough vWF in the blood, or when it does not work the way it should, the blood takes longer to clot.

There are 3 types of vWD.

Type 1 is the most common. With this type, there is less vWF in the blood than normal.

Type 2 is the next most common and includes several sub-types. In Type 2, the vWF does not work properly.

Type 3 vWD is the rarest, and the most severe form. People with Type 3 vWD have almost no vWF and the small amount they have may not work properly. As vWF transports Factor VIII in the bloodstream, people with Type 3 vWD have very low levels of Factor VIII as well. This is another important protein in the clotting process.

Platelet Function Disorders:

Platelet function disorders may affect as many as 1 in 100 women. The majority of these disorders are mild and many women go undiagnosed. However, some types of platelet function disorders, such as Glanzmann Thrombasthenia, are more serious.

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 blood factors can form a clot at the site of an injury.

Other bleeding disorders:

There are a large number of other rarer bleeding disorders both of coagulation factors and of platelets caused by deficiencies of trace proteins in the blood. Men and women can be affected in equal numbers. For more information on your specific disorder please contact your treatment centre, or alternatively visit www.haemophilia.ie for our publication 'Rare Bleeding Disorders' where a number of disorders are explained in more detail.

How are Bleeding Disorders inherited?

Bleeding disorders are generally passed from parent to child at the time of conception. Each cell of the body contains structures called chromosomes. A chromosome is a long chain of chemicals known as DNA. This DNA is arranged into units called genes. These genes determine such things as the colour of a person's eyes. In most factor deficiencies, one of these genes has a mutation. The defective gene that causes a factor deficiency is usually on a chromosome, which does not decide the sex of the child. This means the bleeding disorder has an autosomal inherited pattern and can affect females as well as males. The majority of bleeding disorders have an autosomal inheritance pattern.

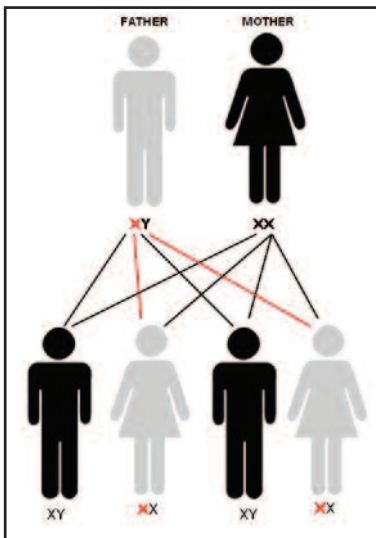


Figure 1: When the father has haemophilia, his daughters will automatically be carriers of haemophilia, but his sons will be unaffected.

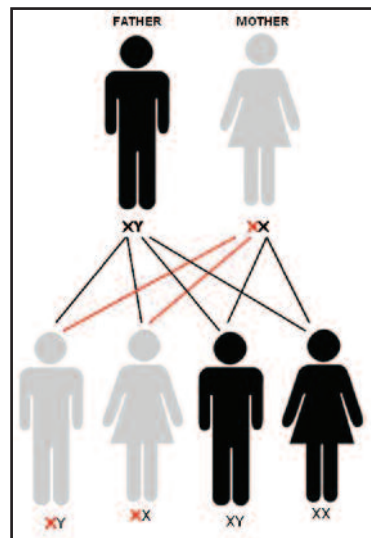


Figure 2: When the mother is a carrier of haemophilia there is a 50 / 50 chance that her sons will have haemophilia and that her daughters will be carriers.

Carrier Testing:

There are two main tests potential carriers must undergo. Firstly, there is a genetic test to determine a woman's carrier status.

Secondly, there is a factor level test to measure her factor levels. This can help determine whether she is at a greater than normal risk of bleeding.

It is important to note that a person with normal factor levels can be a carrier.

Reasons for testing in childhood

As carrier testing is a genetic test, the policy in Ireland is that a girl must be 16 years old before she is tested for carrier status. The sooner a girl is tested after this age, the more time she has to come to terms with being a carrier and all the issues that are associated with it. It also prepares her for eventual decisions such as contraception, pregnancy and childbirth.

It is important to know the factor level in a potential carrier due to the increased risk of bleeding that she may experience with low factor levels, i.e. bleeding after tooth extraction, trauma or surgery. Also when a girl has her period there is a tendency for excessive bleeding and if the girl has very low factor levels she 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.

Reasons for delaying testing until adulthood

There are some arguments against carrier testing at a young age. It may affect a girl's self-esteem and / or can affect the decisions she makes regarding relationships. Some families feel the only reason to know about a carrier status is for family planning.

All girls who are possible carriers should have their factor levels checked in childhood and be referred for genetic testing once they reach 16 years of age. Tests should be carried out at a Comprehensive Care Centre or a Haemophilia Treatment Centre.

The Tests:

When testing carriers of haemophilia A (factor VIII) and haemophilia B (factor IX), the following tests are usually used:

Factor VIII and Factor IX assays: This measures the amount of factor VIII or IX activity in the blood. It is important to note that having a normal factor range does not guarantee non-carrier status.

vWF antigen: This measures the amount of von Willebrand factor in the blood so as to rule out von Willebrand Disease as the cause of bleeding.

Genetic studies: These tests determine the exact genetic mutation and make it easier to provide prenatal diagnosis. There are a number of tests that can be done to determine carrier status. These tests are complex and may take a few weeks to obtain results. Their results are more predictable if DNA from a family member with haemophilia is also available. Obligate carriers or possible carriers of haemophilia should get DNA tested.

Note: Factor levels can be affected by hormone levels.

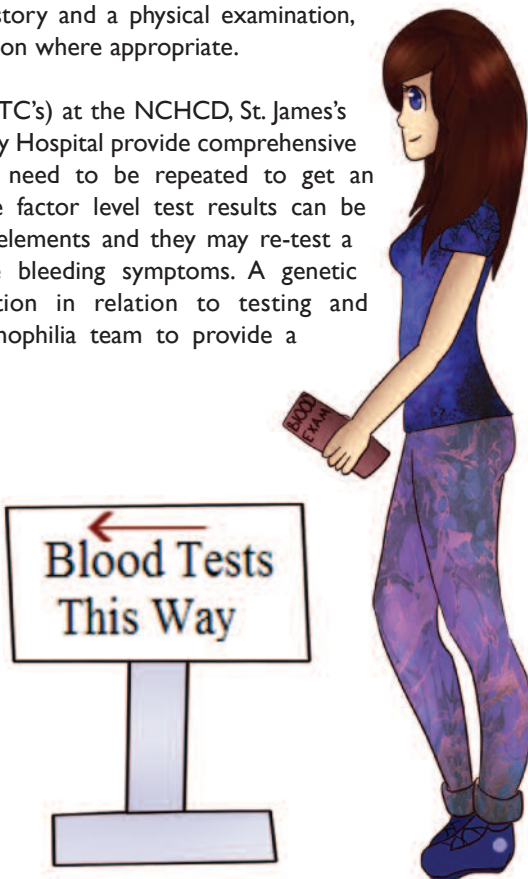
Factor levels in factor VIII carriers may be affected by physical and mental stress, recent use of aspirin or other painkillers, pregnancy, contraceptive pills, exercise, breast-feeding, recent transfusions or infections. So tests may need to be done more than once. Genetic testing for carrier status is best done before pregnancy as factor VIII levels rise during pregnancy, however, testing can be done during pregnancy.

In the case of factor IX carriers, clotting levels are not affected by hormones and testing can take place at any time. Before any levels are checked, patients should have a complete personal and family history and a physical examination, including a gynaecological examination where appropriate.

Haemophilia Treatment Centres (HTC's) at the NCHCD, St. James's Hospital, Dublin and Cork University Hospital provide comprehensive testing. In some cases tests may need to be repeated to get an accurate diagnosis. This is because factor level test results can be affected by a number of different elements and they may re-test a woman if she continues to have bleeding symptoms. A genetic counsellor will provide information in relation to testing and works very closely with the haemophilia team to provide a multidisciplinary approach to care.

Points to remember:

- * A member of your family with or carrying haemophilia must initially give blood samples.
- * The Clinic is co-ordinated by a Clinical Nurse Specialist.
- * A follow up appointment is sent when results are available.
- * A confirmatory sample is requested when attending for results of initial sample.



Signs of Bleeding Disorders in Women

Bleeding disorders can affect men and women. However it can often be more difficult to diagnose bleeding disorders in women.

If heavy periods are something a mother or sibling suffers with, often they are accepted as normal. However, a family history of heavy periods can be an indicator of a family history of a bleeding disorder.

In addition, women with a family history of a bleeding disorder may think their periods are normal because their menstrual bleeding is similar to that of other female family members.



Signs of a bleeding disorder include, but are not limited to:

- * Heavy periods
 - Bleeding for more than seven days
 - Passing clots bigger in size than a 50 cent coin
 - Changing a tampon / sanitary towel every hour or less
 - Flooding
- * Bruising
- * Anaemia
- * Bleeding after sex
- * Nosebleeds
- * Prolonged bleeding from cuts
- * Prolonged bleeding after:
 - Dental work
 - Surgery
 - Childbirth
- * History of muscle or joint bleeds
- * Bleeding when passing stools

If you have any of the signs listed above, make an appointment with your GP. It is important to get a proper diagnosis so that you can receive sufficient treatment, care and support.

Periods



Talking about periods can be embarrassing, but it is important to remember that every woman has periods and knows exactly what it is like to get your first period.

What is a period?

As we grow older our bodies go through puberty, this is when changes occur in the body and our shape begins to change. For women, a part of puberty is menstruation otherwise known as periods. A period is when your body releases tissue it no longer needs. The tissue that is released during a period is the lining of the uterus (womb).

When will I get my period?

Every person is unique so the age at which someone starts their period will differ from person to person. Most young women experience their first period between the ages of 12 and 14 years old. However, it is not uncommon to get your period before the age of 12 or older than 14. However, if you have not started your period by the time you are 16, speak to your mother and / or doctor to make sure everything is ok.

How long will my period last?

Again this is different for everyone. Usually a period will last between 3 to 7 days, but for girls with bleeding disorders they can last longer. The length of time between periods usually ranges from 21 to 28 days, but this varies from person to person. It can take a while for your periods to regulate and for the first year you may be “caught off guard” on occasion, but after a year your periods will begin to come in a regular cycle, which means you should be able to calculate when you will start your period and how long it will last. Talking to your mother and / or sister(s) can help you gauge your cycle.

It is important to speak to someone; a friend, sister, cousin, mother, teacher or doctor if you have any questions or are worried about anything.

First Period

Your first period is a milestone, it marks the start of the transition from childhood into adulthood. However, it is not always an event to celebrate. Advertising can lead girls to believe that they will have a “happy” and “fun” period, but the reality can be quite different.

It is important to know what to expect when you have your first period. Unfortunately many girls are caught off guard for their first period which can be embarrassing. Talking to your mother and sisters about when they started their period will give you an indication of when you may start yours.

Menstruating can lead to several side-effects. Many girls suffer with painful cramps and / or other symptoms such as headaches, moodiness, acne etc. but these can be managed. If you are experiencing pain during your period you should speak to your doctor.

It is estimated that a girl loses 30 to 45 millilitres (2-3 tablespoons) of blood during her period, although the perception is and can feel that you lose more. Women with bleeding disorders usually lose more as they can bleed heavier and / or for longer. Heavy periods, also known as menorrhagia (men-or-ash-e-a), are diagnosed when a blood loss of 80 millilitres (5 ½ tablespoons) or more occurs. It can be difficult to measure how much blood loss occurs during a period, but here are some tips to help you judge the heaviness of your flow:

- * Bleeding for longer than 7 days
- * If you change your tampon or sanitary towel every hour or less
- * Flooding or leaking (soiling clothes)
- * Passing clots the size of a 50cent coin or larger

There can be a lot of information to understand and process about periods, but it is important to know what is normal and what is not. Below is a chart to help you discover if your periods are normal or heavy.

NORMAL PERIOD	MENORRHAGIA
Changing a tampon or sanitary towel several times during the day.	Changing a tampon or sanitary towel every hour.
Occasionally missing activities because of your period.	Missing activities every time you have your period.
Occasionally soiling clothes or underwear with blood.	Regular soiling of clothes or underwear with blood.
Wearing a sanitary towel or tampon at night.	Needing to change your sanitary towel or tampon several times during the night.
Bleeding for up to 7 days.	Bleeding for more than 7 days.

Please note: The above are guidelines and do not define what a period should or should not be like.



Should I use sanitary towels or tampons?

The decision to use a sanitary towel or a tampon is completely up to the individual, both do the same job, but in different ways.

A sanitary towel is an external pad which absorbs blood when it leaves the body. A sanitary towel is worn in a woman's underwear and is secured by an adhesive strip on the back of the pad.

A tampon is an internal plug which absorbs blood before it leaves the body. A tampon is inserted into the vagina and expands when inside to absorb fluids. A tampon has a cord which is visible outside the body for easy removal. Many girls find it painful to use a tampon for the first time, but the pain should go away as you become more comfortable and confident in using this method of protection. If pain persists it may be because the tampon is not inserted correctly, be sure to read the instructions that come with the tampons. If you are struggling to insert a tampon correctly don't worry, that's why instructions are given. Take a deep breath and remember practice makes perfect.

There are side effects associated with using tampons so it is important to read the instructions and guidelines in the pack carefully. A tampon should never be left in for more than 8 hours. Always ensure that you remove your old tampon before inserting a new one. Leaving a tampon in for more than the recommended time can lead to infections or on rare occasions, toxic shock syndrome (TSS). Toxic shock syndrome is an illness caused by a bacteria. A common cause of TSS is leaving a tampon inside the body for more than the recommended time. The string from the tampon should always be outside the body to ensure safe removal. If the string breaks or is internal a tampon can be removed using your hand. If you cannot remove a tampon or are showing signs of TSS visit your doctor immediately.

There are different levels of protection available in both sanitary towels and tampons ranging from light to super absorbent. It can take time to discover what level and type of protection you need so it is important to be patient. Keeping notes on your cycle and flow should help you choose a protection that suits you. It is important to note that flooding and leaking may occur if the level of protection you use is not suitable to your flow, but again this is something that you will figure out as you grow.

Top Tips for Being Prepared for your Period

It can take a year for your cycle to become regular so it is important to be prepared. Here are a few tips to ensure you are never caught off guard:

- * Keep a small make up bag in your school bag or handbag with a spare pair of underwear and some tampons / sanitary towels.
- * Keep a record of your periods to work out your cycle.
- * Insert a new tampon / use a new sanitary towel just before you go to bed so there is less chance of flooding or leaking.
- * Wear dark coloured underwear and trousers / pyjama bottoms so that if an accident does occur the stain will not appear as visible.

Menstruation means more than periods. Many girls and women experience cramping, mood swings, headaches, bloating and many other side effects. Here are some tips to help you cope during the monthly visits!



- * Avoid wearing clothes that are tight – some people can feel bloated when they have their periods and having tight clothes on will make the situation worse and also make you feel self conscious.
- * Use a hot water bottle – a hot water bottle is the next best thing to a hug, so whether you have cramps or are just feeling a little down, fill up your hot water bottle and hold it tight. Just remember to have a cover on; a hot water bottle should never be placed directly onto skin. Also remember the water should be warm, not too hot!
- * Do some exercise – although you may not feel like it, exercise will help relieve tension in the muscles and ease the pain caused by cramps.
- * Have some “me” time – have a warm bath, give yourself a manicure, throw on your favourite tracksuit, have a pyjama day or watch a movie. You may not have a “happy period”, but you can try and what better way to do it than by spoiling yourself – you deserve it!
- * Treat yourself – whether it is a packet of crisps, a bar of chocolate or a take away, satisfy your food cravings!
- * Avoid stressful situations – your periods should not impact on your everyday life, but it can be an emotional time. Don't put yourself under pressure to take part in activities or situations you feel uncomfortable in.

No two people will experience the same period so it is important to remember that nothing you experience is wrong. If you are worried about any symptoms you have, speak to someone about your concerns.

Treatment options

Periods will occur every month, unfortunately there is nothing we can do to stop this! However, for girls who experience menorrhagia there are treatment options that will help manage blood loss.

The Contraceptive Pill

The pill can help regulate periods and make them lighter as it contains synthetic hormones that replicate the properties of oestrogen (east-row-gin) and trick the body into thinking there are higher levels of oestrogen in the blood therefore making the period lighter. This method of treatment is not suitable for anyone trying to have a baby as it is a contraceptive.



As with any medication there can be side-effects from taking the contraceptive pill. Common side effects are weight gain, acne, moodiness or nausea. It is important to consult your doctor before starting any medication and if you experience any side effects, alert your doctor as perhaps another tablet or method of treatment would be better suited to you.



An IUD

An Intrauterine (in-tra-you-t-her-in) device is a small t-shaped device that is positioned in the uterus, the device releases a small amount of the hormone progesterone which helps to reduce the heaviness of your flow. There is no guarantee that your period will become regular with an IUD. This method of treatment is also a contraceptive so should not be used if trying to become pregnant.

Before an IUD can be inserted, a physical examination must take place. This allows the doctor to rule out any infections, but also ensure that the IUD is suitable.

An IUD can be inserted at any time during the menstrual cycle, but it is recommended to have the device inserted immediately after your period finishes. The device must be inserted and removed by a doctor. Tampons can still be used when an IUD is in place.

Desmopressin

Desmopressin is a synthetic drug, a copy of a natural hormone, and is used in the treatment of von Willebrand Disease and occasionally mild haemophilia A. It works by releasing the body's stored factor VIII and / or von Willebrand factor in the bloodstream. Desmopressin must be prescribed by a doctor and can be taken as an injection either into a vein or subcutaneously (under the skin) or as a nasal spray. Some common side effects of desmopressin are cramps, nausea, headaches and blushing.

Tranexamic acid

Tranexamic acid {also known as Cyklokapron (sigh-clo-cap-ron)} is a synthetic drug that can be used to treat and manage prolonged menstrual bleeding as it slows down the body's natural process of breaking down clots which helps reduce blood flow. Tranexamic acid must be prescribed by a doctor and can be taken as a tablet or as an oral solution. Common side effects of tranexamic acid are nausea, tiredness, dizziness and stomach pain.

Factor Concentrates

Factor concentrates are used to treat men and women with bleeding disorders, but may be used in severe cases of menorrhagia. Factor concentrates replace the factors missing in the blood and allow clotting to take place. Factor concentrates are given by injection into the vein.

Endometrial Ablation

Women with bleeding disorders and/or women who suffer with menorrhagia are treated with medication and given advice on lifestyle. In rare cases when medications are not effective endometrial (end-o-meat-real) ablation may be considered. This is where a thin layer of the uterus lining is removed. In most cases this stops the menstrual flow, but in women who suffer with menorrhagia it makes the flow lighter and more manageable.

Endometrial ablation is a procedure whereby an instrument (hysteroscope) is inserted into the vagina, through the cervix and into the womb and the lining is removed using a heat source. This procedure can be done as an outpatient, but women with bleeding disorders may require hospital admittance. As with any procedure there are risks, some common side effects are cramping, nausea, frequent urination and discharge.

This procedure can lead to complications when conceiving so it is important to discuss your concerns and thoughts with your doctor before considering this procedure.

Frequently Asked Questions

How will I know if I suffer with menorrhagia?

There is no check list for what a period should be and your experiences with your period will be different to those of your friends with theirs. Have a look on page 9. If you have several of the symptoms listed under the column “menorrhagia” then you might have heavy periods. However, these are only common symptoms and do not mean you do suffer with menorrhagia. If you are worried speak to your parents or your doctor.



I'm embarrassed to talk to my doctor about my periods?

Your doctor is a professional and is there to help you. You can ask to see the doctor by yourself or bring someone along if it helps you feel more comfortable. Most doctors' surgeries have a nurse who you can make an appointment with. If you do not feel comfortable speaking to a doctor or nurse, speak to a female family member. Every woman goes through a period so she will understand your concerns and help you decide if you need to speak to a doctor. The important thing is that you talk to someone.

Will my everyday routine be affected by my period?

It shouldn't, but sometimes it may be. When you begin your period it may be painful, but with time you will learn to manage and control the pain. You should be able to go to school, hang out with friends and carry on as normal, but if you find that you can't, then you should talk to a doctor as you may require treatment.

Can I wash as normal?

Yes, it is important to keep fresh and clean, but even more so during your period as you can tend to sweat more. In fact most girls feel better after a shower or bath as they feel fresher, more relaxed and the warm water can help ease cramps. Some women may wear a tampon when having a shower or bath, but this is an individual choice.

Can I swim when I have my period?

Yes, using a tampon ensures that you can swim without any fear of leaking as the blood is absorbed by the tampon internally. Tampons should be changed when finished swimming.

A sanitary towel cannot be used for swimming as the pad will absorb the water making it ineffective for absorbing blood.



How often should I change my tampon or sanitary towel?

This depends on the level of your flow and the absorbency of your tampon or sanitary towel. It is recommended that you change your sanitary towel every 2 to 3 hours, if you need to change your sanitary towel more frequently than this you should change to a sanitary towel with a higher absorbency.

For tampons it is recommended to change every 4 to 8 hours. A tampon should never be left in for more than 8 hours. If after 4 hours you find that your tampon is saturated, you should be using a higher absorbency. If you remove the tampon between 5 to 8 hours and can still see white fibres you may need to look at using a lower absorbency tampon.

It is recommended that you use a sanitary towel at least once a day when on your period, rather than constantly using tampons.

Is it normal to feel tired when I have my period?

Unfortunately yes. Periods bring with them cramps, mood swings, headaches and other side effects all of which can make you feel tired. Tiredness can happen regardless of how heavy your periods are. However, if you have heavy periods, your tiredness may be due to the blood loss which can cause anaemia (ah-knee-me-ah) which is a lack of iron in the blood. If you are concerned that you may be anaemic speak to your doctor who may prescribe or suggest iron supplements for you to take.

Although fatigue is common, it should not prevent you doing everyday activities and should not rule your life. It is important to keep up your routine. Exercising can help relieve period pains and help improve your mood and energy levels. It is recommended that you get 8 hours of sleep a night. Although you may be tired, resting for long periods can actually make you feel worse than better.

Why do I have pain in the middle of my cycle?

Part of the menstrual cycle is ovulation (of-you-lay-shone). This is when the egg leaves the ovary. Ovulation occurs once every month, usually two weeks before your period happens. Ovulation can occur without any symptoms, however many women experience pain and discomfort. This pain is not a sign that anything is wrong, but if you are worried, speak to your doctor.

My friends take tablets for pain relief when they have their periods, is it ok for me to take them?

You should always consult your doctor or nurse before taking any pain relief tablets. Women with bleeding disorders should **NEVER** take aspirin or NSAID's (Non-Steroidal Anti-Inflammatory Drugs). Always read the labels before taking any tablets.

Anaemia

Anaemia is a condition caused by a lack of red blood cells. It occurs when the normal balance of red blood cell production and destruction is impaired, resulting in an overall reduction in the number of red blood cells in the body. This can be caused by:

- * Inadequate red cell production
- * Excessive destruction of red blood cells in the spleen
- * Excessive bleeding

Iron deficiency anaemia occurs when there is a lack of iron in the blood. Women with bleeding disorders and women who suffer with menorrhagia are at a higher risk of developing iron deficiency anaemia as they lose a high amount of blood each month.

Symptoms of anaemia are:

- * Fatigue
- * Pale complexion
- * Dizziness
- * Weakness during or following exercise
- * Poor concentration

Anaemia can only be diagnosed by a doctor who will carry out a blood test to measure the iron levels in your blood. If you have several of the symptoms listed above speak to your doctor about having your blood checked.

Iron deficiency anaemia is treated by increasing the iron intake in your diet (eating more red meat, eggs and beans) and / or taking iron supplements. It is important to speak to your doctor before making any changes to your iron intake.



Minor Cuts and Bruises

Cuts & Scratches

A person with a bleeding disorder does not cut more easily, bleed more or faster than normal, they simply bleed for longer.

Cuts and scratches are common for everyone. For people with bleeding disorders, minor cuts and scratches can generally be managed by adding a little pressure for 10 minutes and using a plaster when the bleeding has stopped.

Bruising

People with bleeding disorders have a tendency to bruise more easily than others. Bruises can look unpleasant, but are generally not serious and will fade in a matter of days or weeks. However, as teenagers and as women, we are more self conscious about our looks and any blemishes such as bruises.

Everyone can bruise so it is important not to hide away, but it can be scary if you have several bruises in visible areas. If you are self conscious speak to you parents.



Some tips to coping with bruises are:

- * Using arnica cream.
- * Wearing long sleeved tops, trousers or tights to cover the bruising until it fades.
- * Using tinted moisturisers or make up to reduce the colouring of a bruise.

If you feel that you are bruising more than normal and it is impacting on your lifestyle, speak to a doctor.

For larger bruises, strains, sprains and swellings it is important to use **RICE** – Rest, Ice, Compression and Elevation.

Rest

No strenuous activity until pain has reduced and swelling subsided.

Ice

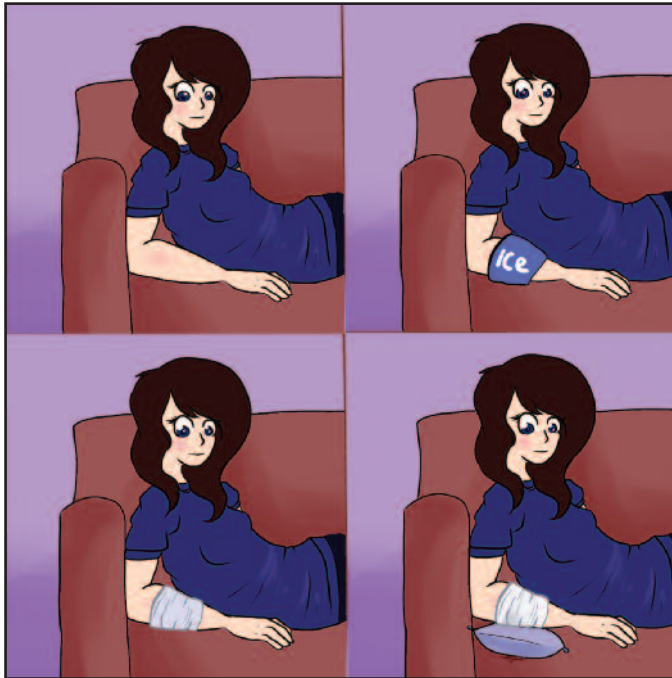
Wrap a cold pack (ice pack or frozen vegetables) in a towel and apply to the effected area for 10 to 15 minutes. Repeat this every two hours.

Compression

When the ice pack is removed, wrap the injured area with an elasticated bandage. Remove the bandage when reapplying ice, going to bed or resting.

Elevation

Raise the injured area above the level of the heart to reduce swelling.



All bleeds and bruising, no matter how big or small, should be dealt with quickly to ensure there is no permanent injury.

If a bruise comes on suddenly, increases in size rapidly and causes pain, treatment may be required. In these instances, inform an adult as soon as possible.

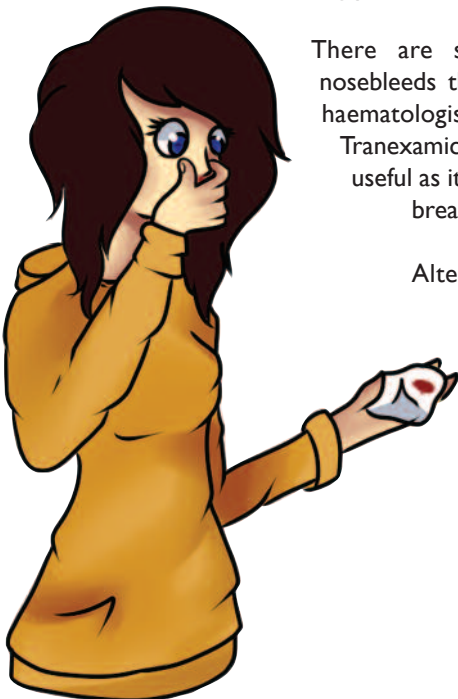
Nosebleeds:

A common and unpleasant symptom of having a bleeding disorder can be frequent nosebleeds (epistaxis).

A nosebleed that occurs for no apparent reason and lasts longer than ten minutes despite pressure on the nose, or a nosebleed that needs medical attention, can be an indication and common symptom of having a bleeding disorder.

Epistaxis is most common in people with severe bleeding disorders. However people with moderate haemophilia, Type 1 and Type 2 vWD should be aware that nosebleeds may be more frequent and last for longer than in a person without a bleeding disorder.

If you get a nosebleed remember to tilt your head forward not backwards and use an ice pack or bag of frozen food on the neck or a cold compress on the bridge of your nose. Use your thumb and index finger to squeeze together the soft portion of your nose. Keep holding your nose until the bleeding stops. Once the bleeding stops, don't do anything that may make it start again, such as bending over or blowing your nose.



There are some medical options for frequent nosebleeds that you may wish to discuss with your haematologist. Your haematologist may prescribe Tranexamic acid tablets (Cyclokapron). These can be useful as it reduces blood flow and slows down the breakdown of clots which are formed.

Alternatively repeated nosebleeds may require some of the blood vessels to be cauterised (sealed by burning) by an ENT (Ear, Nose and Throat) Specialist which is a simple procedure to help seal the blood vessel to stop further bleeding.

Dental

People with bleeding disorders are more likely to bleed during dental procedures so it is important to keep healthy teeth and gums. Making regular visits to the dentist will reduce the chances of future problems such as needing extractions or mouth infections, which can lead to further complications.

If you have mild haemophilia with a factor level of 30% or greater, it is fine to attend your own dentist for routine care such as check-ups, fillings, scale and polish, root canal, crowns and cosmetic work. You should inform your dentist about your bleeding disorder and advise him/her that further information is available from your haemophilia treatment centre. However, should you need a tooth extraction, some deeper injections, gum surgery or a dental implant, your dentist should contact your haemophilia consultant or nurse for advice as this type of treatment will usually need to be provided in a specialist centre where treatment with factor can be given.

Your dentist will advise you about how often you need to attend for check-ups and give you information on how to prevent dental problems. Everyone with haemophilia should attend their dentist at least once a year. Being able to visit a local dentist allows you to get a full range of care in a convenient location. In most cases, once your dentist has more details and guidance about your bleeding disorder from your haemophilia centre he/she will be confident to safely carry out routine dental care.



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.

Gums that bleed after tooth brushing may also be a sign of early gum disease, which is a common problem for everyone. It is important that this does not progress to more severe gum problems. The recognised treatment is to visit your dentist or hygienist for advice and to have your teeth professionally cleaned and monitored. Your dentist may advise you to use a medicated mouthwash and will recommend methods for brushing and cleaning your teeth more effectively.

Extractions should always be co-ordinated by your haemophilia treatment 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 tablets before the tooth is extracted and a special mouthwash for afterwards. In addition the dentist may stitch the wound and use a special surgical packing to stop any bleeding.

Surgery

If you have a diagnosed bleeding disorder you should carry a bleeding disorder alert card or alternatively a medical alert bracelet or necklace in the case of a medical emergency. Any planned surgical procedure should be performed in hospital with a haemophilia treatment centre. Prophylactic (preventative) treatment may be required pre and post-operatively to reduce the risk of excessive bleeding. Girls with inherited bleeding disorders are more likely to have peri-operative (post-surgery) and/or delayed (7-10 days later) bleeding, even with relatively minor procedures.



Sex



In Ireland the legal age of consent to have sex is 17 years of age.

When a girl feels ready to have sex for the first time it can be a nervous time. For girls with bleeding disorders the decision to have sex can come with more worries and concerns. Sex can be an embarrassing subject to talk about – nobody wants to or likes talking to their parents about sex and it is embarrassing speaking to a teacher or doctor. However, it is important to have all the information before taking part in any sexual activity.

Losing your Virginty

Making the decision to have sex for the first time is something that should be thought about carefully. If you are not 100% comfortable with the idea of having sex, then you are not ready. You should never be pressured or coerced into having sex. It is an important moment in your life and you want to enjoy it.

Once you feel you are ready and have made the decision to have sex, you should know the basics:

It might hurt

Although having sex is a pleasurable experience, sex for the first time can be slightly uncomfortable. No matter how confident a girl is in her decision, she will naturally feel nervous and when we are nervous our body tenses. This is what happens to the vagina, it tenses up making penetration more difficult and hence slightly sore. The pain should go away as you become more relaxed. With experience, you will learn what positions feel more comfortable and what works best for you.

You may bleed

Bleeding may or may not occur when you have sex for the first time, but if you do bleed it is important to know that it is normal, it happens to girls whether they have a bleeding disorder or not. This bleeding is caused by the hymen being ruptured during sex. The hymen is a thin membrane covering the vaginal canal. When the penis enters the vagina it must pass through the hymen, which results in bleeding. The level of bleeding that occurs varies for each girl and can range from spotting to heavy bleeding as occurs when you have your period and can last for as little as a few hours up to a few days. Bleeding after sex may occur for the first few times you have sex, but will eventually stop. It is normal to feel tender in your genital region following sex for the first time, but again this will ease and become less frequent the more you have sex.

If you are worried or have any concerns about your health after losing your virginity speak to your doctor.

Remember, sex should be an enjoyable experience for both of you, but it is also a major decision. You should be 100% comfortable with yourself, your partner and your decision. It is important to be prepared so we have come up with some top tips to help you enjoy your experience, but also stay safe:

Talk first

Sex can be spoken about a lot, but not discussed. You may feel comfortable in your decision, but it is important that you speak to your partner and ensure that you both feel the same way. Never assume that because you are ready, your partner is ready.

It is an individual choice whether to tell your partner about your bleeding disorder, but if you are ready to share such an intimate experience surely you can share information about your condition. It may be best for your partner to know about your bleeding disorder so that if you do have a heavy bleed following sex they are not frightened. It may also help them to understand your anxiety. Having a bleeding disorder is nothing to be ashamed about and although it may be an uncomfortable conversation to discuss with your partner, you will feel more relaxed if you know that your partner is aware and understanding.

Preparation

You can never be too prepared! You should never expect your partner to supply contraception, always have protection yourself to ensure that you always practice safe sex. Before engaging in sex you should consult your doctor in relation to contraception.

Lubrication

Losing your virginity can be a nervous time and when we become nervous our muscles can tighten, this can make sex difficult as the vagina becomes tighter and it is harder for the penis to be inserted. Lubrication will make insertion easier and more pleasurable.

Relaxation

Sex should be enjoyable and for this to happen we need to be relaxed. Planning your first time with your partner should include choosing a location, a time that suits you (this is not something that should be rushed) and creating an atmosphere that helps both you and your partner feel ready, relaxed and comfortable.

Aftercare

As mentioned on page 23 you may experience bleeding after having sex for the first time. You should have a spare change of underwear and some sanitary towels / panty liners with you in case you do experience bleeding, this will help avoid any accidents. If bleeding does occur it may be necessary to wash the bed clothes, have clean bedding ready so that any mess can be tidied away. You may be sore in your genital region after sex so try to avoid any strenuous activities in the hours and/or days following.

Talk Again

Losing your virginity is a big step. It is important to talk about what you experienced and how you feel following sex. No matter how you feel, you should speak to your partner as it is something that you both have experienced together and should bring you closer.

The first step you must take before having sex is being comfortable and confident with yourself and your bleeding disorder.

If you have any concerns about sex or about any aspect of your bleeding disorder, speak to your doctor.

Tattoos & Piercings

Body art and modifications such as piercings has increased in popularity, particularly among teenagers and young adults. There are risks associated with tattoos and piercings, but for a person with a bleeding disorder there are more risks to consider.

If you are under 18 you must have parental consent to get a tattoo or piercing.

Piercings

A piercing is a hole created in the body to hang jewellery from for fashion purposes. Many people think of piercings as a harmless act, but any procedure that involves you creating a hole in your body should be thought about carefully and in depth.

Regardless of your bleeding disorder, getting a piercing comes with risks which can include:

*** Infection**

Mouth and nose piercings are the most likely piercings to become infected as there is a high level of bacteria present in these areas.

*** Allergic reactions**

Allergic reactions can be caused by the nickel in the jewellery or the ingredients used in the steriliser and / or cleaning fluids.

*** Scarring**

If a piercing becomes infected or you have an allergic reaction you can be left with significant scarring on the site of the piercing. When you decide to remove a piercing there is a healing time required before the piercing site closes completely.

*** Nerve damage**

If a piercing is not placed correctly it can lead to nerve damage which will damage the tissue surrounding the piercing.

Having a bleeding disorder can further add to these complications as following a piercing a person with a bleeding disorder may bleed for longer, this disrupts the healing process and can lead to a higher risk of infection.

Did you know tongue, cheek and lip piercings can result in gum problems and a tongue piercing can damage the enamel on a person's teeth?

It is important to note that certain body parts take longer to heal than others, for example it can take up to a year for a belly button to heal properly. This means that you are susceptible to infections for up to a year after the piercing is removed.

Tattoos:

Unlike a piercing, which can be removed, a tattoo is for life and is a big commitment. You should think carefully before getting a tattoo to ensure it is the right decision for you.

People with bleeding disorders are discouraged from getting tattoos because of the medical risks associated. If a person with a bleeding disorder makes the decision to get a tattoo it is vital that they speak to the haemophilia treatment centre in advance to ensure they take the necessary precautions.

Time

The size and detail included in the tattoo will determine how long the process will take, tattoos can take several sittings which involve several hours for each sitting. A large tattoo will mean a longer process, resulting in a longer healing time. Everybody bleeds when getting a tattoo as the skin is injected by needles. However, a person with a bleeding disorder may bleed for longer following a tattoo and could suffer with swelling and bruising that can affect healing and also the final look of the tattoo. The time it takes to have a tattoo completed should also be considered as the position of the tattoo could mean posture is affected for several hours at a time.

Infection

As a person with a bleeding disorder will bleed for longer following a tattoo, their healing time will be longer which means they will be more prone to infections. Infections in tattoos can lead to scabbing, which can affect the final turn out of a tattoo.

Before getting a piercing or tattoo you should:

Think about the decision

Once you have made the decision to go ahead and get a piercing or a tattoo. For tattoos you should think carefully about the design, detail and placement of the artwork. The larger or more detailed the piece, the longer the tattoo will take. A tattoo is for life so make sure you are 100% confident in your decision.

Talk to your parents

Depending on your age you may need parental consent. It is best that your parents are aware of your intent to get a piercing or a tattoo so that they can assist you with the aftercare or if any complications arise, they can bring you to the haemophilia treatment centre.

Do your research

There is a high risk of infection associated with tattoos and piercings so it is important to know that the tattoo parlour / piercing shop you intend to use is hygienic and responsible. Visit the parlour / shop before having any procedures

carried out, research them online for reviews and ratings and if possible speak to someone who has used them in the past.

Talk to the Haemophilia Treatment Centre (HTC)

It is important the the HTC are aware of your plans in case any problems arise, they can also give you advice on how to manage your bleeding disorder. Tattoos are not recommended for people with bleeding disorders, but if a person with a bleeding disorder decides to go ahead with a tattoo it is important that they take the necessary precautions outlined by the HTC.

Inform the Piercer / Tattoo Artist

You should inform the person carrying out your piercing of your bleeding disorder status, so they are aware of any potential complications. When consenting to a tattoo most forms will have a notice asking if you have a bleeding disorder. It is important to be honest and inform the tattoo artist of your condition prior to getting your tattoo. If you do not inform the tattoo artist and a problem arises it may result in the tattoo not being completed or not turning out as designed. With prior knowledge the tattoo artist can plan how to carry out the procedure.

Aftercare

Follow instructions given to you by the piercer / tattoo artist and the HTC. It may be your body, but it is their business and they know what is best.

CONSULT YOUR HAEMOPHILIA TREATMENT CENTRE BEFORE HAVING ANY TATTOOS OR BODY MODIFICATIONS CARRIED OUT.



Personal Care and your Bleeding Disorder



Hair Removal

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

Shaving can easily cause anyone to cut themselves, for a person with a bleeding disorder these small cuts can be more inconvenient and take longer to stop. Ways to avoid these mishaps are to avoid using cheap or old razors - if using a disposable razor, replace 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 do nick yourself when shaving, you should allow yourself plenty of time to stop the bleeding before leaving the house.

Waxing can be painful whether home waxing or having a professional carry out the treatment. 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 are using is suitable for your skin. Waxing may cause small blood blisters to appear 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 does occur.

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 methods of hair removal, but results last longer. It is important to notify the beauty therapist of your condition before having laser hair removal carried out as blistering of the skin may occur.

Sports

Exercise and fitness should be part of your everyday routine, regardless of your bleeding disorder status. However, it is important to understand that because of your conditions some sports are deemed unsuitable because of the high risk of injury and bleeding. Exercise is not limited to sports and can include dance and gym sessions. The same precautions should be taken when undertaking any fitness activity. Speak to the haemophilia treatment centre before undertaking any strenuous exercise or joining any sports club to ensure that the activity is suitable for you and your condition. The centre staff can also help you prepare for activities by advising you of what protection, if any, is needed for your chosen fitness plan.

School Plan

School may not be your favourite place, but it is an important part of life. Having a bleeding disorder may result in you missing days from school, this can be due to periods or muscle / joint bleeds. It is important that the school is aware of your bleeding disorder so they can help you to keep up to date with school work and ensure that you do not fall behind. It is also important that they know in case a situation arises when you are at school and need medical attention. Talk to your parents first and decide on what action to take when informing the school. You may decide that you would like your parents to talk to your principal or year tutor or that you would like to be involved on the discussion. Informing the teaching staff does not mean that the pupils will know about your bleeding disorder, telling your fellow pupils and friends is something that you and you only can do. The teaching staff should respect your wishes to keep the status of your condition private.

Pain Relief

You should always consult your doctor or nurse before taking any pain relief tablets. Women with bleeding disorders should **NEVER** take aspirin or NSAID's (Non - Steroidal Anti-Inflammatory Drugs). Always read the labels before taking any tablets.

Dealing with your disorder

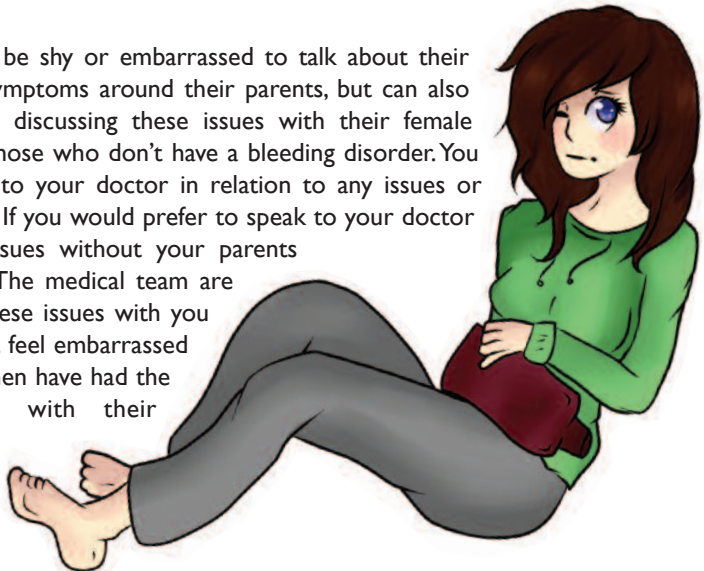
The diagnosis of a bleeding disorder can greatly impact on both the physical and emotional quality of life for a person. Women with bleeding disorders, including symptomatic carriers, are recognised as having a medical condition. However, in the past this was not the case and women often felt they had to deal with their condition alone. Today, there are support services and information available for women with bleeding disorders and carriers to help you deal with your diagnosis.

Part of becoming independent in relation to your bleeding disorder is to understand the impact your diagnosis has on you and your life and learn how to deal with that impact. How severely you are affected by your symptoms will generally determine how significantly your life may be affected by your bleeding disorder. However, most discussions about bleeding disorders focus on the physical consequences, but the emotional effect shouldn't be downplayed or ignored.

At the time of diagnosis with a bleeding disorder, you may experience a large variety of emotions – shock, denial, anger, frustration and self-blame. You may be afraid of what it will mean for you, your relationships and any children you may have. You may feel a sense of loss, that you are different from everyone else and may even fall into a pattern of denial and refuse to acknowledge the diagnosis.

These emotions may be particularly heightened if there is a history of complications due to bleeding disorders in your family. It is important to acknowledge these emotions as a normal process and to talk to someone in confidence about how you are feeling.

Young women can be shy or embarrassed to talk about their bodies and their symptoms around their parents, but can also feel uncomfortable discussing these issues with their female friends, especially those who don't have a bleeding disorder. You should try to talk to your doctor in relation to any issues or concerns you have. If you would prefer to speak to your doctor about particular issues without your parents this can be arranged. The medical team are there to discuss these issues with you and you should not feel embarrassed – many young women have had the same discussions with their doctor.



It may also help to recognise the following:

You're not alone

If you're going through something difficult at the moment, there's a good chance that someone else has, and probably is, experiencing the same thing right now.

Your emotions and feelings are very real

Pay attention to them; you can work through them. You'll be much better off in the long run.

Accepting and managing your emotions are not easy or quick processes

That said, the quicker you accept and take charge of your emotions and feelings, the quicker you'll feel better about life, and the easier it will be to deal with those emotions.

Create a network of friends who will support you

Find someone you can talk to; an adult, your doctor or a peer. It may be someone who gives advice or it may be just someone who will sit and listen.

Get out and do something

When you are down, it can seem like too much effort to get ready and do an activity with friends but it may be just what you need to lift your spirits.

If this does not work

Talk to someone you trust, there is a team in the haemophilia treatment centre who are ready and willing to help you deal with any issues you may have due to your diagnosis.

The tips for management of your bleeding disorder discussed in this publication and with your haemophilia treatment centre can help you to feel a sense of control and think positively about your bleeding disorder diagnosis.



For support, information and advice on any issues covered in this booklet contact:



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