



THE
HAEMOPHILIA
SOCIETY



Understanding
von Willebrand disease

Contents

Introduction.....	3
What is von Willebrand disease?.....	4
What causes von Willebrand disease?.....	5
Types of von Willebrand disease	7
Diagnosing von Willebrand disease.....	10
Treating von Willebrand disease.....	12
Living with von Willebrand disease.....	16
Women and von Willebrand disease.....	19
General guidance.....	21
Glossary.....	22
Our services	23

Introduction

This booklet is for anyone who has been diagnosed with a bleeding disorder called von Willebrand disease (VWD). It gives a general introduction to VWD and information on diagnosing, treating and living with the condition. You may also find the information useful if you are concerned that you might have a bleeding disorder.

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

"For most of my life having a bleeding disorder has NOT been a problem; it has not stopped me doing anything." Chris



What is von Willebrand disease?

von Willebrand disease (VWD) is a bleeding disorder. It is a common inherited condition that can sometimes cause heavy bleeding. It is caused by a deficiency of von Willebrand factor (VWF), which is a type of protein that helps your blood to clot. von Willebrand disease is different from haemophilia, another type of bleeding disorder.

It is the most common inherited bleeding disorder, possibly affecting thousands of people in the UK. It affects males and females equally.

There is currently no cure for VWD but for most people it does not usually cause serious problems, as the condition is mild and manageable. Most people with it can live normal, active lives. However, the lower the level of VWF, the more likely you are to have bleeding problems. In the most severe form the body does not make any VWF at all.

VWD affects the blood's ability to clot. If your blood does not clot you can bleed more than most people and have symptoms such as easy bruising, frequent or long-lasting nosebleeds or bleeding from your gums. Women are more likely to experience symptoms and complications due to the increased risk of bleeding during menstruation, pregnancy and childbirth. It may also be hard to stop bleeding after an injury, dental procedure or surgery.

A protein in the blood called von Willebrand factor (VWF) helps blood to clot. In VWD, either the level of VWF is low or the VWF does not work very well, or both. Normally, when a blood vessel is injured you start to bleed. Small blood cells called platelets clump together to plug the hole in the blood vessel and stop the bleeding. For most people, VWF acts like glue to help the platelets stick together and form a blood clot. When you have VWD the glue does not stick the platelets together, so clots do not form as easily.

VWF also carries a blood-clotting factor called factor VIII (eight), another important protein that helps your blood to clot. Factor VIII is the protein that is missing or does not work well in people who have haemophilia, another bleeding disorder. Some people with VWD also have low levels of factor VIII and may share some symptoms with people with haemophilia.

What causes von Willebrand disease?

VWD is almost always inherited. This means it is passed from parent to child through their genes. Genes are the instructions that control our growth and how our bodies work. VWD can occur when the gene that contains instructions for producing the VWF protein is altered. This is sometimes known as a fault or mutation.

You can inherit type 1, 2 and pseudo-VWD, which are all passed on in an autosomal dominant manner. This means that only one of your parents passes a faulty VWF gene on to you. For each pregnancy there is a one in two chance that the child will have VWD.

You can inherit type 3 VWD if both your parents pass a faulty VWF gene on to you (autosomal recessive manner). Your symptoms may be quite different from your parent or parents' symptoms.

There is also an 'acquired' type of VWD that can develop later in life, often as a result of another condition. This is not covered in this booklet.

In some cases, you can have a faulty VWF gene without any symptoms of VWD. However, you can still pass the faulty VWF gene on to your children.

The type of VWD a person is born with mostly depends on whether they inherit copies of this faulty gene from one or both parents.

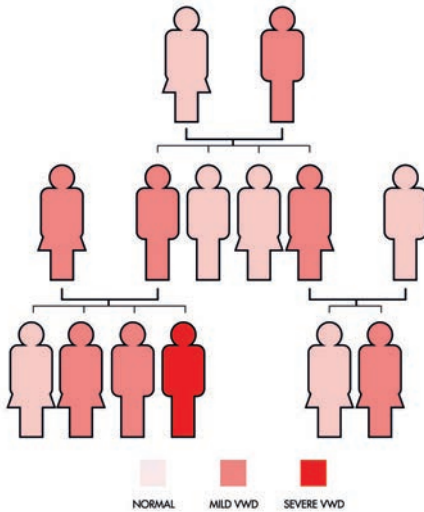
- **If one parent has a genetic fault that causes VWD** – there is a 1 in 2 (50%) chance of a child having type 1 or type 2 VWD, and no chance of them having type 3 VWD.
- **If both parents have a fault in the gene** – there is a 1 in 2 (50%) chance of a child being a carrier like their parents and a 1 in 4 (25%) chance of them having type 3 VWD.

The chances of inheriting mild forms of type 1 VWD can also be affected by other things, including blood group: people with blood group O are more often affected than people with blood group A or B.

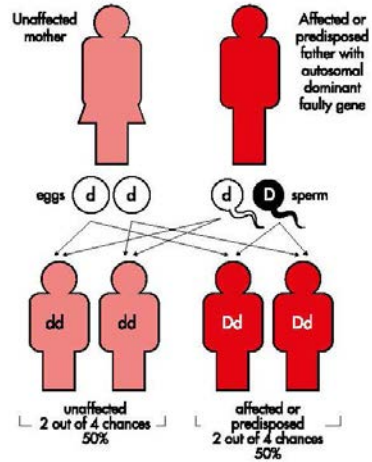
This means it is not always easy to predict whether a child might inherit it from their parents.

Parents who are carriers of a faulty VWF gene may not have symptoms themselves.

Inheritance of von Willebrand disease



Autosomal Dominance Inheritance



Low von Willebrand factor

Some people have low VWF levels that may contribute to bleeding but are not low enough to be called VWD. This applies to people with levels of VWF between 30-50% of normal.

In general people with low VWF levels (30-50% VWF activity) do not have a faulty VWF gene. VWF levels can be reduced for other reasons, such as when the body clears VWF from the blood much faster as happens in some people who have blood group O.

If you have low VWF levels and have bleeding symptoms like people with VWD you may need treatment if you are having a tooth taken out or an operation.

Interpretation of VWF test results can be challenging and will need consultation with a doctor who specialises in bleeding disorders, such as a haematologist. You may need to have more than one blood test to make a clear diagnosis.

Types of von Willebrand disease

The three major types of VWD are called type 1, type 2 and type 3. Different gene faults cause each type. The severity of symptoms depends partly on the type of VWD a person has. Types 1 and 2 are generally mild, but people with type 3 VWD can have very serious bleeding episodes. Even within each type of VWD, symptoms can be quite variable.

Type 1

Type 1 is the common form of VWD and is generally the least severe. About 3 in 4 people who have VWD have type 1.

People who have type 1 VWD have low levels of VWF and may have low levels of factor VIII. Bleeding is mostly only a problem if people have particularly low VWF levels, have surgery, injure themselves, or have a tooth removed.

Type 2

In people with this type of VWD, von Willebrand factor does not work properly. Bleeding tends to be more frequent and heavier than in type 1. About 1 in 4 people who have VWD have type 2.

There may be a low level of VWF and sometimes factor VIII is low too. The important difference is that the quality of the VWF is affected. This means it is less effective in helping the blood to clot.



There are four main subtypes of type 2 VWD. These are: 2A, 2B, 2M and 2N.

- **Type 2A**

The VWF multimers are not the right size.

- **Type 2B**

The VWF multimers are not the right size and the VWF becomes too active. It attaches to the platelets in the blood when it is not supposed to. The body quickly gets rid of the platelets with the attached VWF. This causes a shortage of both platelets and VWF in the blood.

- **Type 2M**

Low or absent binding to receptor on platelets. VWF is not able to stick to the platelets. Factor VIII binds as normal.

- **Type 2N (Normandy)**

Autosomal recessive. Factor VIII levels reduced to 5-25%, as VWF has a reduced affinity for factor VIII.

Type 3

This is the most severe and rarest type, affecting about 1 in 500,000 people.

People who have type 3 VWD have virtually no VWF. If you have this type, your body will not produce any VWF. As a result, your platelets will not be able to form a clot and your FVIII level will be low as well. This will put you at risk of severe bleeding that is difficult to stop. Bleeding from the mouth, nose and gut is common, and joint and muscle bleeds can occur after an injury.

Symptoms

The symptoms of VWD depend on which type you have, although everyone is affected differently. Many people have such mild symptoms in day-to-day life that they do not know they have VWD. Others have frequent and heavy bleeding that affects their everyday life and need regular treatment to manage their condition.

If you have had a diagnosis of VWD, it is important to speak to your haemophilia centre if you are bleeding. If you are not sure if you have VWD but have one or more of the symptoms listed below it is important you see your GP. It may be helpful to take this booklet with you.

Type 1 or type 2 VWD or low VWF

When you have VWD bleeding can stop and start over a number of days. It may not seem to be a lot at any one time, but it can be excessive over several days. Possible symptoms include:

- frequent or large bruises from minor bumps or injuries, particularly on your arms and legs
- frequent or hard-to-stop nosebleeds
- heavy, delayed or prolonged bleeding after surgery
- prolonged bleeding from gums after dental procedures
- heavy bleeding from a cut
- prolonged or heavy bleeding after an injury
- bleeding gums
- heavy or prolonged menstrual periods (menorrhagia)
- bleeding during or after labour
- spontaneous bleeding, e.g. internal or joint bleeding (only in the most severe of cases).

Type 3 VWD

People who have type 3 VWD may have any of the symptoms listed above. They may also have bleeding into muscles or joints. Bleeding may happen for no obvious reason.

Diagnosing von Willebrand disease

Because symptoms can be mild, VWD can be difficult to diagnose and often goes undetected. Most GPs will not have met anyone with VWD so getting a diagnosis can take persistence and repeated visits. Your diagnosis will usually take place in a Haemophilia Comprehensive Care Centre or a Haemophilia Treatment Centre, where you will see a haematologist (doctor specialising in blood disorders) or a specialist haemophilia nurse.

VWD is often hard to diagnose. People with type 1 or type 2 VWD may not have day-to-day bleeding problems. This means it is quite common for people not to be diagnosed until they have heavy bleeding after surgery, dental treatment or injury. For women, giving birth may be the first time that VWD is suspected.

Type 3 VWD can cause major bleeding problems in babies and children. This means people with type 3 VWD are usually diagnosed during their childhood.

Medical history

Your doctor or specialist nurse will ask you questions about your personal and medical history, and your family's medical history of abnormal bruising and bleeding. This helps your medical team understand how your VWD affects you and can be helpful in diagnosing you correctly. It can be useful to keep a diary of your bleeding and bruising to show the doctor, who may ask about any:

- bruising with little or no injury, especially if you could feel a lump under the bruise
- nosebleeds that happened for no reason and lasted more than 10 minutes despite pressure on the nose, or any nosebleeds that needed medical attention
- bleeding from a small wound that lasted more than 15 minutes
- prolonged, heavy or repeated bleeding that needed medical care after having a tooth out or an operation
- heavy and/or prolonged monthly periods
- medicines you have taken that might cause bleeding or increase the risk of bleeding (e.g. aspirin and non-steroidal anti-inflammatory drugs (NSAIDs) such as ibuprofen).

Type 3 tends to be the easiest to diagnose. If you have it, you are likely to have a history of severe bleeding starting early in life.

Physical examination

Your doctor may examine you to look for any unusual bruising or recent bleeding.

Blood tests

No single test can diagnose VWD. Diagnostic testing to confirm VWD may have to be repeated because levels of VWF fluctuate and having these tests more than once to confirm or exclude a diagnosis is vital. Anxiety, exercise, pregnancy, illness and stress can also have an effect, so it is not unusual to get slightly different results each time.

Your consultant will need to collect a sample of your blood, which they will send to a laboratory for testing. Due to the specialised nature of these tests, it may take up to two or three weeks to get your results.

Many people need multiple tests over a period of time to make a clear diagnosis, including which type of VWD they have. Blood tests may include:

- von Willebrand factor antigen: this test measures the amount of VWF in the blood
- von Willebrand factor ristocetin cofactor activity and/or collagen binding activity: these tests show how well VWF works
- factor VIII level: this is checked because if VWF is low, factor VIII level may also be low
- blood group.

Your consultant will also be able to test for other bleeding disorders, which will form part of the differential diagnosis.

'Being told that I had a bleeding disorder was actually a relief as I had begun to think I was imagining it.' Chris

Family screening

After the diagnosis of VWD has been made, testing should be offered to first-degree relatives (with or without a positive bleeding history).



Treating von Willebrand disease

Minor bleeds such as nosebleeds, bruises and minor cuts can often be controlled using simple first aid measures.

Treatment may be needed for having teeth out, surgery or after an injury. It is important you contact your haemophilia centre or comprehensive care centre for advice before treatment if you:

- need to have a tooth out
- need surgery
- are injured
- are pregnant.

Treatment options include Desmopressin (DDAVP®), VWF replacement therapy and tranexamic acid. A new treatment approved by the European Commission is Veyvondi®.

Desmopressin (DDAVP®)

DDAVP® is a synthetic drug modelled on a hormone found normally in the body. It is not used in children under two years old as it can cause fluid retention that is hard to control. DDAVP will usually stop bleeding in people with mild to moderate VWD with episodes of spontaneous or trauma induced injuries such as bleeds into a joint (haemarthroses), bleeds into a muscle (intramuscular haematomas) or bleeds from the soft tissues like the nose or gums (mucosal bleeding).

How does it work?

DDAVP® stimulates release of your own clotting factors (factor VIII and VWF) from storage sites in the body into the blood. Levels of the clotting factors are increased by three to six times your baseline level for 12-24 hours. If necessary, you may have a repeat dose after 12 hours. In some people repeated infusions may not be as effective because the body doesn't have the chance to rebuild its stores. The dose is calculated according to body weight.

DDAVP® trial

As some people do not respond to DDAVP® you will need to have a trial infusion. This is to see how well you respond to the treatment before it is needed for a bleeding episode or an emergency.

You are given a dose of DDAVP® and then blood samples are taken at set time points after the injection for up to six hours. Occasionally a blood test will be needed on the following day. This helps the doctors understand how your body responds to the drug.

The response to DDAVP® will be reported as one of the three following categories:

- DDAVP® responsive: DDAVP® is the treatment of choice for all procedures and bleeding episodes except major surgery or injury.
- Partial response: DDAVP® can be used only for minor procedures and minor bleeding episodes. An alternative treatment will be necessary for major procedures and major bleeding episodes.
- No response: an alternative treatment will be necessary for all procedures and all bleeding episodes.

How is DDAVP® given?

DDAVP® can be given:

- by intravenous infusion (a drip into a vein) over 30 minutes to an hour
- as an injection under the skin (tummies and thighs can be good places for this; the needles are similar to those used by people with diabetes)
- in a nasal spray. (Always use Desmopressin nasal spray exactly as your doctor has told you.)

Your centre will advise which method is best for you.

Before you have DDAVP®

It's important you tell your nurse or doctor the following before you have DDAVP®:

- if you have any known reaction to this or any other medication
- if you take diuretics (drugs that increase urine production), medication for high blood pressure or any other medicines, including over-the-counter medicines and herbal remedies
- if you are pregnant or breastfeeding
- if you have any significant medical problems including heart disease, kidney disease, cystic fibrosis, epilepsy.

Side effects of DDAVP®

Occasional side effects of DDAVP® include:

- headache
- facial flushing
- stomach pain and nausea (feeling sick)
- allergic reactions
- decrease in blood pressure
- increase in heart rate.

Treatment with DDAVP® without reducing fluid intake may lead to fluid retention (see page 14) and dilution of salt in the blood. Very occasionally, in more severe cases it can lead to epileptic seizures. If these side effects occur, your doctor may advise an alternative treatment to DDAVP®.

Fluid intake

As DDAVP® can cause the body to retain fluid you will be asked to restrict fluid intake after having the drug. Your haemophilia centre will advise you on how much you should drink.

Factor VIII/VWF

Factor VIII/VWF concentrate is given into a vein to replace the missing VWF and allow clotting to take place. It is made from donated pooled human plasma (the fluid part of blood). Most people in the UK will be on a plasma-derived product, though the first recombinant treatment is available here. It is pure VWF and does not contain FVIII. It is an injectable drug to control bleeding in adults (age 18 and older) with VWD when DDAVP® treatment alone is ineffective.

This treatment may be used if you:

- cannot take DDAVP® or need extended treatment
- have type 1 VWD that does not respond to DDAVP®
- have certain subtypes of type 2 VWD
- have type 3 VWD.

Tranexamic acid

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.

A liquid form is also available for children, though this usually has to be obtained 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 VWF replacement therapy. Tranexamic acid does not help to form a clot. This means it cannot be used instead of DDAVP® or factor VIII/VWF 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 (loose, watery stools)
- stomach pain.

Dental procedure treatment options will depend on the type of VWD you have and the severity. A treatment plan prior to any dental procedures will focus on your bleeding management strategies. Your dentist will liaise with your haemophilia centre to discuss how to minimise a bleed during and after dental procedures.

Following dental extractions your dentist may use oxidized cellulose and stitches to aid clotting.

Factor replacement therapy for people with moderate and severe forms of VWD can ensure certain dental treatments can be done safely. Treatment should take place as soon as possible after the administration of factor concentrate – normally within 30 minutes to an hour.



Living with von Willebrand disease

'Others don't understand how much we have to consider in everyday life to work around treatment, work arrangements, trips to friends and holidays. It's easy to feel isolated. However, I am now part of The Haemophilia Society's women's group. This has definitely helped me to feel less isolated.' Clare

Tell your doctor, dentist, pharmacist and other health professionals that you have VWD. Many health professionals will not know anything about VWD as it is a rare disease, so having some information to give them or asking them to contact your haemophilia centre is a good idea. Your dentist can ask your haemophilia centre whether you need treatment before dental work to reduce bleeding.

You also may want to tell other people about your condition, like an occupational health nurse or sports coach. Anyone who cares for a child with VWD (e.g. teachers, childminders, nursery staff) should be told about the condition. This will allow them to act quickly and appropriately if you or your child has an injury. It can be helpful to explain that you have a full understanding of your or your child's VWD and are competent in managing it in daily life.

'I just want to live life and be normal. I hate having bleeds. When one starts, my heart sinks and I think, oh no, not again!' Sam

Consider wearing a medical ID bracelet or necklace if you have a serious form of VWD (e.g. type 3). In case of a serious accident or injury, the healthcare team treating you will then know straightaway that you have VWD.

Non-prescription medicines

Avoid over-the-counter medicines that can affect blood clotting, such as aspirin, ibuprofen and other non-steroidal anti-inflammatory drugs (NSAIDs). Always check with your haemophilia specialist before taking any medicines.

Exercise, fitness and sport

It is a really good idea to keep fit and active. The types of exercise that work well for people with bleeding disorders include swimming, cycling and walking. High contact sports such as rugby and boxing are not safe activities if you have bleeding problems. Always check with your haemophilia centre before starting any exercise programme but don't let your bleeding disorder stop you being active.

'I have type 2 VWD and despite having chronic or occasional GI bleeding for the last seven years, I consider myself fit and well. My exercise regime consists of Tai Chi for one hour three times a week followed by a 30-minute swim.' Jane

Dental care

It is important that you take good care of your teeth and gums to reduce the chance of future problems such as extractions or mouth infections, which can lead to further problems. You should visit your dentist at least twice a year for advice on interventions such as fluoride treatments and fissure sealants (special coatings on the teeth) to prevent dental problems.

Travel

If you are planning to travel it is important to find out the contact details of haemophilia centres in the places you are visiting. Information is available from your haemophilia centre, the World Federation of Hemophilia (wfh.org), or you can call us on 020 7939 0780. Make sure you take up-to-date written medical information with you, including your diagnosis and the name and phone number of your haemophilia centre.

'I try and make sure our VWD doesn't stop my daughter and me from doing anything and travel is no exception. We don't need regular treatment so I just make sure I know where the nearest haemophilia treatment centre is and that if we go abroad, we have great travel insurance; I know general policies can't cover us, but it's easily sorted. Oh, and I always have an ice pack for all the bumps and bruises she gets having fun.' Kelly

It is very important to have travel insurance that covers your VWD. You may find our travel insurance factsheet helpful; it also has lots of other top travel tips. You can find it on our website or call us on 020 7939 0780 for a copy.

Tips for travellers

- Always carry your own treatment products, treatment equipment, and pharmaceutical drugs with you, as products will be different and probably very expensive in another countries. Most countries also have only limited supplies.
- Carry a letter to present to customs, if requested, to explain why you are carrying treatment products, prescribed drugs, needles and syringes etc.
- When travelling by air, always carry your own treatment products, treatment equipment and pharmaceutical drugs on the aircraft as hand baggage. This will allow you to present them quickly to security and customs, if requested. There are risks of loss, breakage, and of temperature variations potentially affecting treatment products if put in the aircraft hold.
- Carry a letter from your treating doctor with information about your bleeding condition, any blood-borne viruses you may have, and the usual treatment you receive.



Women and von Willebrand disease

Periods

It can sometimes be difficult for a woman to tell if her periods are heavy. Comparing yourself to other women in your family can be misleading, as they may also have VWD without knowing it.

These signs may be useful in assessing a potential problem:

- Bleeding which lasts longer than seven days.
- Bleeding that affects daily life, such as bleeding so heavily that you don't want to leave the house or worry about flooding and it showing through clothes.
- Tiredness and lethargy (lack of energy) or shortness of breath due to anaemia (lack of red blood cells) caused by heavy bleeding.

If you have heavy periods you might want to carry extra sanitary protection or even a change of clothes in case of accidents. There is no need to just accept that you have heavy periods and adapt your life to fit around them, as treatment is available. Talk to your haemophilia centre as oral contraceptives, other hormone therapy and anti-fibrinolytic drugs can all be effective.

'As a teenager I had very heavy periods until I went to university, when my new GP suggested that I went on the pill as it would help to control the bleeding – it worked.' Chris

Pregnancy and childbirth

You can have children if you have VWD, even if it is severe, although there is risk of your child being born with VWD and/or you having heavy bleeding during or after labour. Talk to your doctor about your bleeding disorder before becoming pregnant. With proper care, you can have a successful pregnancy and deliver a healthy child.

As soon as you know you are pregnant, contact your haemophilia centre. You should be monitored closely throughout your pregnancy by your obstetrician (doctor specialising in pregnancy and childbirth) under the guidance of your haemophilia centre. Working together in this way will help the doctor who delivers your baby take special safety measures to avoid injury. These safety measures include not using forceps or vacuum extraction (Ventouse Suction) to assist in the delivery of your baby, if possible.

Pregnancy can cause blood levels of VWF to increase, reducing the likelihood of bleeding complications during pregnancy and delivery. However, this needs to be monitored as women with VWD can have heavy bleeding/haemorrhage after giving birth or have delayed bleeding when the factor levels return to pre-pregnancy levels. This can happen any time from 24 hours after birth to six weeks after delivery.

It is important for the doctors and midwives looking after you to be aware of your VWD; not just for your sake but for your baby's too. It should be assumed that the baby may have VWD and delivery methods should be as gentle as possible. It is also important that everyone knows about your VWD if you need a caesarean delivery. Women with VWD whose VWF does not rise to normal levels during pregnancy will need specialist assessment and multidisciplinary team management.

If you continue to bleed heavily during the days and weeks after the birth, you should remind your midwife or health visitor about your VWD and seek treatment. Remember that most healthcare professionals may be very unfamiliar with VWD. Always let your haemophilia centre know if you are pregnant or are experiencing bleeding so that they can advise the team looking after you about the risks for you and your baby and how best to care for you both. It is important not to presume your maternity team have contacted your haemophilia team, even if they're based in the same hospital, as this can sometimes be missed.



General guidance

If you have VWD, it is a good idea to:

- Tell your surgeon or dentist about your VWD if you are due to have a procedure or operation – you may need to take medication to reduce the risk of bleeding before and afterwards. Your surgeon or dentist may also need to contact your doctor to discuss the procedure.
- Tell your doctor or nurse about your VWD if you need a vaccination – they can give the injection just under your skin to avoid painful bleeding in your muscles.
- Avoid aspirin and non-steroidal anti-inflammatory drugs (NSAIDs) like ibuprofen unless your specialist advises you it is safe to use them, as these can make bleeding worse – use other medicines such as paracetamol instead.
- Ask your haemophilia doctor or nurse if there are any activities you need to avoid – you should be able to take part in most sports and activities, but it is best to check first.
- Ask your haemophilia centre to provide you with a Bleeding States Information Card – you always need to keep this card with you so if you are involved in an emergency you can show the card and give attending staff the relevant information and phone numbers to call for advice.
- Boys at risk of VWD should **not** be circumcised without first speaking to your Haemophilia Centre, due to the risk of excessive bleeding.

If you prefer, you can buy a MedicAlert disc or piece of jewellery. As you wear them, they are a good way of making sure your medical details are always to hand. Visit www.medicalert.org.uk to find out more.

Glossary

Acquired

Acquired bleeding is not inherited, or passed through families, like most bleeding disorders.

Collagen binding activity

Measures the quantity of von Willebrand factor molecules bound to collagen in the blood vessel wall.

Comprehensive care

A way of giving health care in which a team of professionals works with the patient to improve his or her physical, emotional, and mental wellbeing. The care is usually given in one place; either at a haemophilia comprehensive care centre (HCCC) or haemophilia treatment centre (HTC), so the experts can work together.

Factor concentrate

A factor protein that has been made into a powder.

Factor VIII (Factor 8)

One of the clotting factors that is essential to prevent bleeding. The clotting factor protein that is decreased in people with haemophilia A.

Fibrin

Strands of protein that weave around and through a platelet plug to form a blood clot.

Fissure Sealant

A protective layer that keeps food and bacteria from getting stuck in the tiny grooves in the teeth.

Haemophilia

A lifelong hereditary blood disorder in which bleeding lasts longer than normal. It is caused by a defect in a protein needed for blood clotting.

Hereditary

Passed in the genes from parent to child. The basic unit of heredity is the gene.

Joint

The place where two or more bones come together.

Menstrual period

Also called menstruation or just a 'period'. The shedding of the lining of the uterus through the vagina. On average, it occurs every 28 days and lasts from three to five days.

Multimer

Formed by many von Willebrand factor protein units joined together.

Platelet

A small disc-shaped particle in the blood that is used in the clotting process. Also called a thrombocyte.

Platelet function tests

Determine how well the platelets work.

Pseudo or platelet-type

Is similar to type 2B, but the defect is in the platelets instead of in the factor.

Ristocetin cofactor activity

This test is used to evaluate the function of the protein von Willebrand factor (vWF), which helps blood to clot.

Our Services

The Haemophilia Society – For everyone affected by a bleeding disorder

We are the only UK-wide charity for all those affected by a genetic bleeding disorder; a community of individuals and families, healthcare professionals and supporters.

For most 70 years we have campaigned for better treatment, been a source of information and support, and raised the awareness of bleeding disorders.

We want to ensure that everyone affected by a bleeding disorder:

- Lives the best life that they can
- Never feels alone or isolated
- Feels empowered and confident

We do this by:

- Raising awareness about bleeding disorders
- Providing support at all life stages
- Influencing and advocating on policy and access to treatment

More than 30,000 men, women and children in the UK have a diagnosed bleeding disorder, and the number rises every year. Membership of The Haemophilia Society is free and open to all.

To find out more, or to become a member for free, visit our website at haemophilia.org.uk or call us on **020 7939 0780**.



THE HAEMOPHILIA SOCIETY

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

We hope you have found this information helpful. If you have any comments or suggestions about this booklet or any of our other information, please contact us via the email below.

Your Society: getting in touch

The Haemophilia Society

Phone: 020 7939 0780

Email: info@haemophilia.org.uk

Web: haemophilia.org.uk

 [HaemophiliaSocietyUK](https://www.facebook.com/HaemophiliaSocietyUK)

 [HaemoSocUK](https://twitter.com/HaemoSocUK)

 [thehaemophiliasociety](https://www.instagram.com/thehaemophiliasociety)

Registered charity no. 288260 (Scotland SC039732)

Company limited by guarantee reg. no. 1763614

Members of the European Haemophilia Consortium and the World Federation of Hemophilia

President: Baroness Meacher

© The Haemophilia Society 2019

Review date January 2021