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
von Willebrand disease (VWD) is the most common inherited bleeding disorder, possibly affecting thousands of people in the UK. It affects males and females equally.

VWD affects the blood’s ability to clot. If your blood doesn’t clot you can bleed more than most people and have symptoms such as easy bruising or nosebleeds, and women may have heavy periods. It may also be hard to stop bleeding after an injury 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 doesn’t 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 doesn’t stick the platelets together, so clots don’t 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 doesn’t work well in people who have haemophilia, another bleeding disorder. Some people with VWD also have low levels of factor VIII and may share the same symptoms as people with haemophilia.
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 permanently altered. This is sometimes known as a fault or mutation.

You can inherit type 1 or type 2 VWD if only one of your parents passes a faulty VWF gene on to you. You inherit type 3 VWD if both your parents pass a faulty VWF gene on to you. Your symptoms may be quite different from your parent or parents’ symptoms.

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.
Low von Willebrand factor

Some people have low VWF levels which may contribute to bleeding but aren’t 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) don’t 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.
The three major types of VWD are called type 1, type 2 and type 3. Different gene faults cause each type.

**Type 1**
People who have type 1 VWD have low levels of VWF and may have low levels of factor VIII. Type 1 is the common form of VWD. About 3 in 4 people who have VWD have type 1.

**Type 2**
There are four main subtypes of type 2 VWD. These are: 2A, 2B, 2M and 2N. About 1 in 4 people who have VWD have type 2.

In type 2 VWD 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’s less effective in helping the blood to clot.

**Type 3**
People who have type 3 VWD have virtually no VWF. As a result they also have very low levels of factor VIII. Type 3 is the most serious form of VWD, but it is very rare, affecting about 1 in 500,000 people.

**Acquired von Willebrand disease**
Some people get VWD later in life as a result of other medical conditions such as heart disease or some cancers. This type of VWD is called acquired von Willebrand disease (AVWD).

**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 don’t 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’s important to speak to your haemophilia centre if you are bleeding. If you aren’t sure if you have VWD but have one or more of the symptoms listed below it’s 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 nose bleeds
- heavy or prolonged periods
- heavy, delayed or prolonged bleeding after surgery and childbirth
- prolonged bleeding from gums after dental procedures
- heavy bleeding from a cut
- prolonged or heavy bleeding after an injury.

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.
Most GPs won’t 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’s 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 questions about your medical history and your family’s medical history. 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 required medical care after having a tooth out or an operation
- heavy and/or prolonged monthly periods
- medicines you’ve taken that might cause bleeding or increase the risk of bleeding (e.g. aspirin and non-steroidal anti-inflammatory drugs (NSAIDs) such as ibuprofen).
Physical examination
Your doctor may examine you to look for any unusual bruising or recent bleeding.

Blood tests
No single test can diagnose VWD. 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.

You will have these tests more than once to confirm or exclude a diagnosis. This is because small things like how the blood sample is taken, the temperature it is stored at and the time of day it’s taken can all affect the results. Anxiety and stress can also have an effect, so it isn’t unusual to get slightly different results each time.

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

Chris
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’s 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, tranexamic acid and fibrin glue.

**Desmopressin (DDAVP®)**

DDAVP® is a synthetic drug identical to a hormone found normally in the body. It isn’t used in children under two years old as it can cause fluid imbalance that is hard to control.

**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 don’t respond to DDAVP® you will need to have a trial infusion. This is to see how well you respond to the treatment before it’s 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:

1. **DDAVP® responsive**: DDAVP® is the treatment of choice for all procedures and bleeding episodes except major surgery or injury.

2. **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.

3. **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.

Your centre will advise which method is best for you.

**Before you have DDAVP®**

It is important you tell your nurse or doctor the following:

- Any known reaction to this or any other medication
- If you take diuretics, medication for high blood pressure or any other medications, including over-the-counter medications and herbal remedies
- If you are pregnant or breastfeeding
- 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 below) 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).

This treatment may be used if you:

- can’t take DDAVP® or need extended treatment
- have type 1 VWD that doesn’t 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’s often used to prevent or treat bleeding from mucous membranes such as the inside of the mouth, nose, gut or womb. It’s usually taken as tablets, three or four times a day, but may also be given as an intravenous infusion (drip into a vein). A liquid form is also available for children, though this usually has to be obtained from haemophilia centres.

Tranexamic acid may be given before dental treatment, for nosebleeds or prolonged or heavy periods. It may be used alone or in combination with DDAVP® and VWF replacement therapy. Tranexamic acid doesn’t help to form a clot. This means it can’t 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.

Fibrin glue

Fibrin glue is made from synthetic copies of two proteins normally found in the body - fibrinogen and thrombin. Fibrin glue can be put directly onto the site of bleeding. It’s especially useful in tooth extractions.
‘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 won’t know anything about VWD as it’s 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, 11

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 medications

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 sports

It’s 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 aren’t 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 7 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

Travel

If you’re planning to travel it’s 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.

It’s 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 at haemophilia.org.uk/travel or call us on 020 7939 0780 for a copy.
'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
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 your clothes.
- Tiredness and lethargy (lack of energy) or shortness of breath due to anaemia (lack of red blood cells) caused by heavy bleeding.

There’s 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.

Pregnancy and childbirth

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 post delivery.

It’s 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’s also important that everyone knows about your VWD if you need a caesarean delivery.

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 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’s 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.
‘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 and it worked’

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

For 65 years, we have campaigned for better treatment, been a source of practical information and advice, and enabled people living with long-term conditions to:

- lead fulfilling lives
- make informed choices about their treatment and care
- support and inspire others to do the same.

More than 25,000 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.

We bring people together at events like children’s activity weekends, information days for women who have a bleeding disorder, and weekends for families with a newly diagnosed child – giving them the knowledge and support they need to feel confident about the future.

Our peer support through local groups around the UK, global family network, and online community offers friendship and a listening ear when needed, as well as enabling people to share their views and experiences. We also support people experiencing particular difficulties or feelings of isolation, such as developing inhibitors which stops their treatment from working, or experiencing a loss of independence as they grow older.

As a health charity, we work alongside the NHS to:

- provide easy access to information and opportunities
- influence national policy and practice to make the care and treatment of bleeding disorders consistent, effective and accessible to all, and
- enable the voices of all people with bleeding disorders to be heard.

As bleeding disorders are fairly rare, many people will never encounter The Haemophilia Society; we are largely invisible outside of the communities we serve. So we have to work doubly hard to raise both awareness and understanding of bleeding disorders and vital funds needed to give those affected the services they deserve and need to live life well.

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 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 write to the Head of Membership and Planning at the address overleaf.