CSL Behring
Biotherapies for Life™

von Willebrand disease

A guide for school staff and teachers

How to use this guide

This booklet has been written as a guide for teachers and school staff who have a young person with von Willebrand disease (VWD) – a bleeding disorder – in their care.

It explains what VWD is, how it affects people and how to recognise the signs and symptoms of a bleed. It also provides a place to record information about the young person's care.

While schools are not expected to manage or administer medication for VWD, key staff should be able to identify when treatment may be needed. Schools should develop a care plan with the young person's parent or guardian, to ensure appropriate action is taken.

All adults who come into contact with the young person with VWD at school should read this guide. It will help them to develop a basic understanding of the disorder and how to support the young person in the school environment.

Contents

Top five things to remember

If you are involved in the care of a young person with VWD, the main things to remember are:

1. In VWD, bleeding must be treated as soon as possible, with first aid and/or with medication when necessary
2. Pay attention and take action if the child/young person reports pain or the sensation of a bleed or is displaying any other signs of a bleed
3. You can’t always see bleeding; it can be internal
4. Treat the young person as a normal pupil and involve them fully in school life
5. However, avoid full contact sports, unless otherwise agreed

About von Willebrand disease

Von Willebrand disease (VWD) is the most common inherited bleeding disorder, affecting up to 1 in 100 people.

VWD occurs when people lack a substance called von Willebrand factor (VWF) in their blood, or if it doesn't work properly.

VWF is an essential component for blood clotting, and without it, blood is unable to clot normally. That's why, when people with VWD bleed, it takes longer to stop.

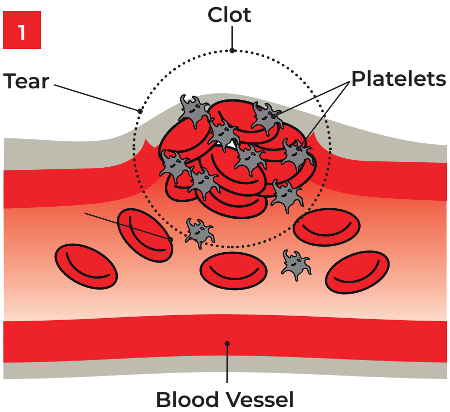
When we bleed from a cut, small blood cells called platelets clump together in a clot to plug the injury. Normally, VWF acts like a glue to stick the platelets together and strengthen the clot. *(See illustration 1)*

But in VWD, this 'glue' effect is reduced, so clots take longer to form or they are weak. *(See illustration 2)*

VWD can affect both boys and girls. It is usually inherited, when one or both parents pass a faulty VWF gene to their children.

It is impossible to 'catch' VWD of any type. No-one at your school needs to worry about it being passed on like a cold or flu.

1



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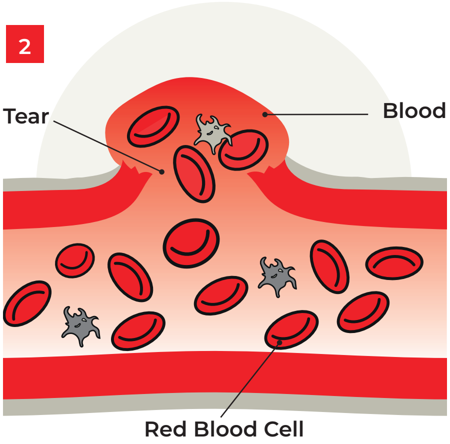
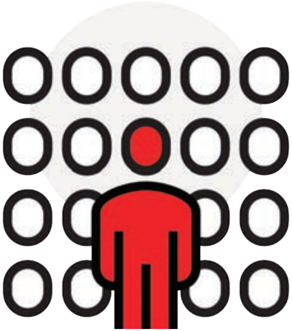




VWD affects 1 in 100 people

Symptoms

The type of VWD that the young person has will affect the severity of their symptoms. Some of the common symptoms include:

- frequent hard-to-stop nose bleeds
- bruising more easily, and having significant bruising from minor bumps
- bleeding for longer than most people after injuries, surgery or dental procedures
- heavy or prolonged periods

In severe cases of VWD, internal bleeding – such as in the gut, muscle or into joints – can occur in addition to all or some of the above symptoms. This can happen even without an obvious trigger, like a fall or bump to the body.

IMPORTANT: Central lines

Any child with a central line that has signs of a high temperature must have an immediate medical review to ensure that there is no infection around the device. Please contact the parents/care giver to arrange this.

Treatment for von Willebrand disease

Anyone diagnosed as having VWD is under the care of specialists at a haemophilia centre. The school should keep the centre's contact details on record (for instance, in the back of this booklet).

Parents/guardians quickly become experts in their child's condition and treatment, and they will usually be your first stop for any questions or concerns.

A stock of the young person's medication should be stored securely at the school, in case of a bleed that cannot be stopped with first aid. Treatment is usually administered by a parent/guardian, or – in secondary school – by the young person themselves.

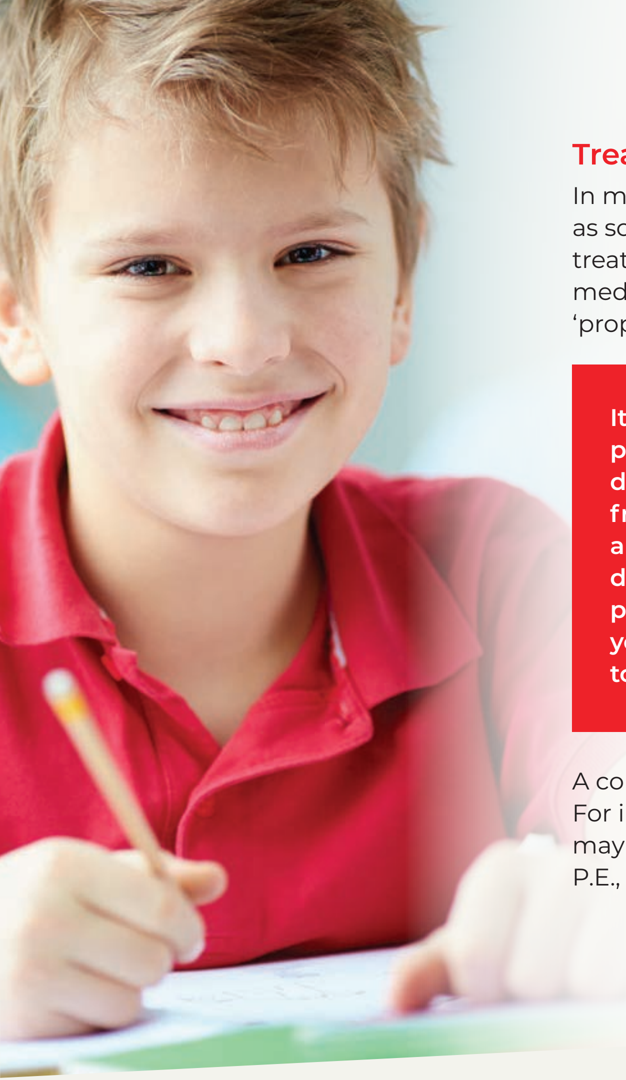
Types of treatment

There are different treatments available to stop prolonged bleeding in VWD. Treatment given depends on type and severity of VWD along with response to medication.

Treatments may be taken orally as a tablet or liquid, via a nasal spray, injected under the skin, or given intravenously (into a vein).

Intravenous treatments are either injected into a vein in the arm, or via a 'central line'. This is a catheter or port that connects directly to a major vein in the chest, and tends to be fitted for weeks or months at a time.

Other than oral medication which schools can often support a patient to receive, all other treatments would only be given by a medical professional, the parent/guardian or young person if trained to do so.

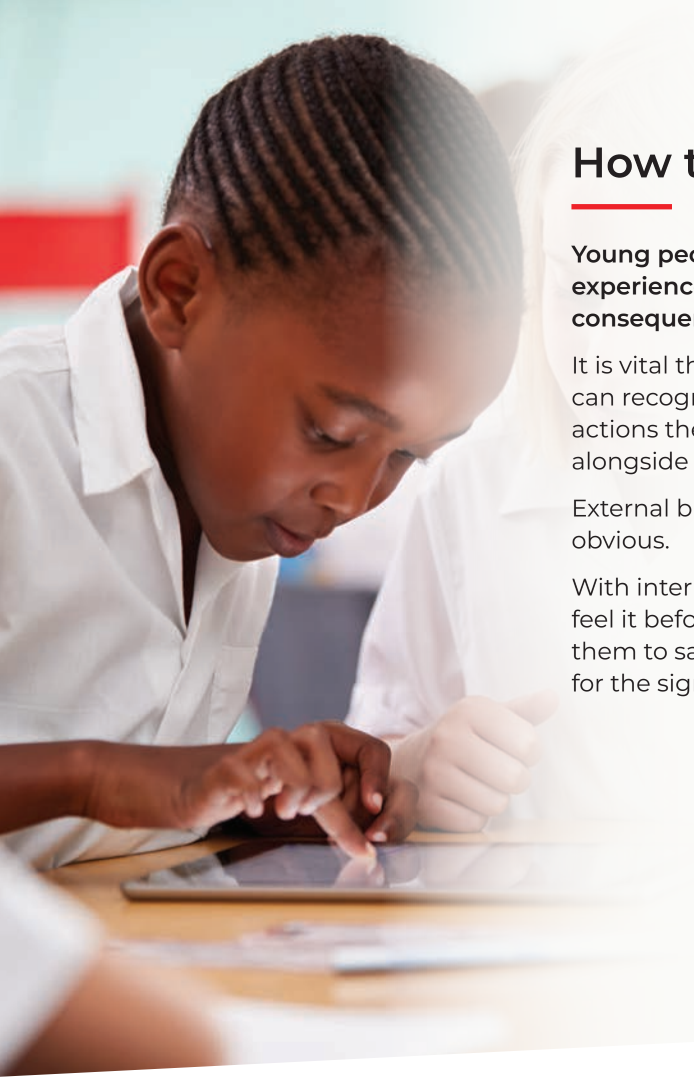


Treatment regimes

In many young people, VWD is managed by treating only when needed, as soon as possible after bleeding occurs. This is known as ‘on demand’ treatment. However, young people with severe symptoms may take medicines regularly, to prevent spontaneous bleeds. This is known as ‘prophylactic treatment’ or ‘prophylaxis’.

It is important to be aware that young people using prophylactic treatment don’t have the same level of protection from bleeds at all times. Treatments give a boost to blood clotting, which then diminishes over time. Discuss with the parent/guardian, to find out when the young person may be more vulnerable to bleeds.

A combination of prophylaxis and on demand treatment may also be used. For instance, young people that normally rely upon treatment on demand may also take a prophylactic approach on higher-risk days, such as before P.E., to provide protection.



How to recognise a bleed, and what to do

Young people with VWD can bleed for longer than normal and can experience internal bleeding. This can cause short and long term consequences if not addressed and/or treated rapidly.

It is vital that school staff who have contact with a young person with VWD can recognise the signs and symptoms of a bleed, and are confident in the actions they need to take. The school's first aid policies should be followed alongside the young person's agreed care plan.

External bleeding – for instance, from a cut or from a nosebleed – may be obvious.

With internal bleeding in the muscles or joints, the young person can often feel it before it is outwardly obvious. That's why it's important to encourage them to say if they think they are having a bleed, and for adults to look out for the signs.

Recognising a bleed

section to be completed by school as part of care plan

Type of bleed	Notes and what to look out for	Action required
Minor cuts	• Monitor for prolonged bleeding	
Bruises	• If bruise is large, swollen and painful it could indicate an internal bleed	
Nosebleeds	• Look out for prolonged or restarting bleeding	
Mouth/gum bleeds	• These can be difficult to stop as blood clots can be dislodged or washed away by saliva, food, drink, or the tongue	
Joint or muscle bleeds	• Most likely in people with severe VWD, but can happen in anyone with VWD after an impact • Can cause long term damage without prompt treatment • The young person may report a tingling or bubbling feeling, or pain, in their limb • Look out for swelling, tight skin, warm to the touch. Not wanting to use/bend/put weight on a limb • Young children may be tearful or grumpy without apparent reason, or you'll notice they're not using a limb • Older children may not want to report a problem. Are they acting out of character, limping or looking uncomfortable?	

Continued on next page

Recognising a bleed

section to be completed by school as part of care plan

Type of bleed	Notes and what to look out for	Action required
Abdominal bleeds	• Most likely in people with severe VWD, but can happen in anyone with VWD after an impact • Keep a close eye on the young person with VWD if they've had an impact to the tummy, side, or lower back • Look out for paleness, pain and vomiting of blood • In pre-schoolers, notice blood in bowel movements or urine. School-age children should be encouraged to report this themselves	
Head, face, neck or throat bleeds	• Head injuries aren't always obvious straight away. Look out for headache, nausea/vomiting, blurred vision, dizziness, sleepiness, confusion, limb weakness • Throat bleeds can cause difficulty breathing or swallowing	

GENERAL POINTS

- Treat serious injury or illness just as you would in any other pupil, by calling 999 and the parent/guardian. Tell the emergency services that they have VWD, tell them if you have treatment on the premises, and notify the young person's haemophilia centre
- School staff can support a patient to receive oral medication as part of an agreed care plan but will not be expected to administer other treatments for VWD. This will be done by the parent/guardian or the young person themselves, if able
- In young people who have a central line, a temperature could indicate an infection associated with their catheter or port. Contact the parent/guardian
- Following any bleed, clear up spills and decontaminate surfaces according to normal school procedures

School life

Young people with VWD can enjoy school as much as any other. The condition does not affect their capabilities, and teachers should concentrate on what they can do, and involve them fully.

- Make sure parents/guardians can easily access the school during the day in case they need to administer medication
- Agree where and how medication will be stored safely and securely at the school
- Ensure teachers know if the young person likes to talk about their personal experience of VWD, or not
- Agree a strategy for catching up on schoolwork if hospital appointments or severe bleeds lead to time off school
- Plan ahead to ensure the young person can safely participate in school trips (see page 13)

Sports

Being physically active is important for health and wellbeing, and should be encouraged in young people with VWD. As well as the importance of them feeling included, improved strength and fitness may help to reduce the frequency of bleeds.

The suitability of particular sports needs to be considered according to the severity of the young person’s disorder, and should be discussed with their parent/guardian who will have guidance from the child's clinican. Full contact sports, such as rugby and martial arts, are not recommended.

School trips and events

Young people with VWD should be included in extra-curricular activities and school trips. When planning activities away from the school, consider the following:

- Who will be responsible for carrying medication, and how/where it will be stored
- Who will be responsible for carrying the young person's bleeding disorders card
- Where the nearest haemophilia centre is, and their contact details
- Who will administer treatment if it is needed. Can the child/young person self administer? Or will a parent need to accompany them or arrange to attend?

Menstruation

VWD can cause heavy and prolonged periods in young women. This can cause anxiety and embarrassment if she frequently has to leave classes to change her pad or tampon. She may also worry about bleeding through into her clothes.

Teaching staff should be conscious of this, and afford privacy and understanding.

Information record

Complete these details to provide a quick reference for staff involved in the care of the young person with VWD. Keep this alongside a copy of the young person's latest clinic letter, and a copy of their bleeding disorders card.

		Updated
Young person's name and DOB		
Name and contact details for first-line contact (parent/ guardian)		
Name and contact details for second-line contact (parent/ guardian)		
Haemophilia centre contact details		
Name of consultants		
Date completed/ updated	____/____/________	____/____/________

Information record

Complete these details to provide a quick reference for staff involved in the care of the young person with VWD. Keep this alongside a copy of the young person's latest clinic letter, and a copy of their bleeding disorders card.

		Updated
Name of nurse specialists		
Names of people who can administer treatment		
Name of treatment		
Where and how treatment is stored at school		
Words young person had used to describe the feeling of a bleed		
Date completed/ updated	____/____/________	____/____/________

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