Introduction

This leaflet aims to provide you with general information about the signs and symptoms your child may have when they have Type 1 VWD. If you are ever worried about your child please contact your IBD Team or take your child to the Accident and Emergency Department.

What is VWD?

Type 1 von Willebrands Disease (VWD) is a genetic disorder of the clotting (coagulation) system in which there is a reduction in the quantity of the von Willebrand protein (VWP) or von Willbrand Factor (VWF). This protein is one of many clotting factors that work in combination to form a stable clot at the site of injury. People affected by VWD bleed for longer but not faster than other people.

There are three main types of VWD

- **Type 1** is found in between 60-80% of people with VWD. This is the reduction in the amount of VWF.
- **Type 2** is found in between 15-30% of people with VWD. This is the reduction in quality of VWF. The levels may be almost normal but the protein does not work properly. Type 2 VWD is split into further classifications type 2A, type 2B, type 2M and type 2N. Symptoms can be mild to moderate.
- **Type 3** is found in between 5-10% of people with VWD this is the reduction or absence of VWF. Symptoms can be more severe.

How is the diagnosis made?

The diagnosis is made by taking a “bleeding history”. You will be asked if your child bruises or bleeds easily and about problems with any surgery or dental procedures. If your child is very young you may not be able to answer some of these questions and we understand this. You will also be asked if other family members are affected by any bleeding or bruising.

As well as asking you questions we will take some blood to check how the blood is clotting and to see if the VWP is low. Sometimes these bloods will have to be repeated as the levels can fluctuate. Further blood tests including genetic bloods are often taken to confirm the type of VWD and to help us to provide you with accurate information and provide the most effective treatment for your child. Genetic tests can only be undertaken with consent.

Where did my child’s VWD come from?

VWD is commonly inherited, passed from one parent who is affected by VWD to their child. However less commonly a spontaneous genetic error can occur and your child may be the first person in the family to have VWD.
Once there is genetic confirmation of your child’s disorder we will be able to advise you if you or other family members need to attend for investigations. We can arrange for the necessary genetic tests to be carried out at Alder Hey or you can be seen by a geneticist.

Children with type 1VWD have few day to day problems relating to VWD but may require some treatment for injuries, surgery and dental extractions. The most common symptoms in VWD are bleeding from the nose and mouth (mucosal bleeding).
How does blood clot?

Clotting factors are proteins in the blood that control bleeding.

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

At the same time, chemical signals are released from small sacs inside the platelets that attract other cells to the area and make them clump together to form what is called a platelet plug.

On the surface of these activated platelets, many different clotting factors work together in a series of complex chemical reactions (known as the coagulation cascade) to form a fibrin clot. The clot acts like a mesh to stop the bleeding.

Coagulation factors circulate in the blood in an inactive form. When a blood vessel is injured, the coagulation cascade is initiated and each coagulation factor is activated in a specific order to lead to the formation of the blood clot. Where one or more of the clotting factors is reduced or not working properly the coagulation cascade is affected slowing the time for the fibrin clot to form.

What does Type 1 VWD mean for day to day life?

Although it is tempting, you do not need to "wrap your child up in cotton wool". You can get on with day to day baby and child play as well as almost all activities for children. We do advise against some contact sports such as rugby, judo and karate.

What are the signs and symptoms of type 1 VWD?

- Bruising
- Prolonged bleeding post dental extractions
- Prolonged bleeding post-surgery
- Bruising or bleeding after vaccinations
- Nosebleeds
- Bleeding gums
- Heavy periods
How should you ask for advice?

If you are worried about your child please try to contact the IBD Team on the numbers provided. However if it is out of hours, and you have been unsuccessful, please do not wait at home for a response but attend the Accident and Emergency Department. Make sure you tell them on arrival that your child has Type 1 VWD.

Situations when you should ask for advice

If your child:

• has a significant head injury with any signs of the following signs:
  o Babies: any signs of high pitched crying increased sleepiness decreased alertness, reduced feeding or vomiting.
  o Older children: any signs of visual disturbances, loss of consciousness, severe or ongoing headache, loss of coordination, tiredness and decreased alertness, difficulty in thinking or severe or persistent vomiting
• has a prolonged nosebleed which does not seem to be slowing after 10-15 minutes has a loose tooth or bleeding from the gum that is hard to manage
• has a heavy difficult to manage period

Please let us know if your child needs surgery or dental extractions. It is important for us to know so that we can make a simple plan to make sure the procedure runs smoothly.

Can my child join in with sports?

Yes, most sports are encouraged as they can help to make your child’s muscles stronger. Contact sports such as rugby, mixed martial arts and boxing should be avoided, tag rugby can be participated in. Other sports including football, swimming, dancing, basketball and cycling are encouraged and your child should be allowed to join in.

Treatments for type 1 VWD

Use usual first aid for nose and mouth bleeds. Apply pressure to the affected area, for example, where a tooth has fallen out, a finger wrapped in gauze pressing firmly over the socket or for nose bleeds firm pressure below the bridge of the nose. Sucking on an ice pop helps both nose and mouth bleeds to slow down as it cools the area.

Injuries and heavy bruising use normal first aid measures first; this includes Protect Rest Ice Compression Elevation otherwise known as PRICE

Protect and (pain killers) - Make sure your child is in a safe place and that the injured or painful area is not at risk of being knocked, paracetamol may be needed to help the pain to settle

Rest - Rest the affected joint or muscle. If your son has an elbow or shoulder bleed he should rest his arm not use it to lift or carry things. If it is a joint or muscle bleed in his leg he should not walk but rest as much as possible.

Ice - Apply ice, wrapped in a towel, or a cold pack to the affected joint either by using an ice pack, crushed ice in plastic bag or frozen vegetables, apply for 10-15 minutes. Ice helps to reduce pain and muscle spasms as well as reducing swelling and redness. Ice alternating with heat can also help to reduce pain, particularly when the bleed is into a muscle.

Compression - Applying pressure to the injured area will help slow blood flow; the pressure can also provide comfort. A Tubigrip bandage is a good example of compression and can be used over an ankle, knee, elbow or wrist.. Compression is often too painful in the early stages of a bleed.
Elevation - Raise the injured arm or leg above the heart although this may seem difficult you can prop an arm and leg up with cushions or pillows. This helps to reduce the blood flow to the area, lower blood pressure to slow the bleeding.

Currently there is no cure for VWD but there are medicines and treatments available that help to keep symptoms under control

**Tranexamic acid** is helpful for gum bleeds, nose bleeds, dental extractions and periods. It works by helping to stabilise blood clots while the blood vessel repairs itself. It is usually given orally but may be given as an infusion intravenously.

**DDAVP** (Desmopressin) an injection that goes just under the skin (subcutaneously). It is used, during surgery or dental work and sometimes for nose bleeds and heavy periods. It works by releasing VWF and Factor 8 from the lining of the blood vessels raising the levels to near normal levels. It cannot be used for children under 2 years old and does not work for everyone. We will arrange for a Desmopressin trial to see if it will help us to manage your child’s von Willebrand disease.

**VWF** is given as an injection into the vein. It is used during surgery or dental work and sometimes for nose bleeds that do not stop. It works by increasing the VWF in the blood stream bringing the factor levels up to normal so that the bleeding will stop. Many children with type 1 VWD do not need this treatment as tranexamic acid and desmopressin work well enough for them.

**Who do I tell?**

It is helpful for other healthcare professionals to know including your GP and Health Visitor.

Child minder, play group, nursery and schools need to know as do the leaders of any club or group your child joins. We routinely visit schools to talk to teachers and lunchtime staff and provide them with written information.

We might advise you to talk to family members who may need to ask a doctor to review if they could be affected by VWD.

You might find it helpful to tell friends about VWD so that they can support you and so that if your child develops bruises while playing with their children they can understand why.

**How often will we see the IBD Team?**

We will see you or speak to you as often as you would like. We know that lots of questions or worries do not wait for a clinic appointments.

Routinely we will see you with your child at diagnosis and then in clinic for primary immunisations, we give all immunisations just under the skin. This is a good time for us to get to know you and for you to get to know the team. It is a good time for you to ask any questions and for us to talk a little about VWD. After this we will see you in clinic first at 3 and then 6 month intervals. By the time your child is five years old we will see you routinely once a year. You might feel happier coming more often this can be easily arranged.

**Medicines that should be avoided:**

Please avoid using pain killers known as NSAIDs including ibuprofen, naproxen and diclofenac as these interfere with the way the blood clots by affecting the function of platelets, NSAIDs can also irritate the lining of the stomach, causing bleeding. You can give your child paracetamol if he needs a painkiller or something to reduce a temperature.
Links

The Haemophilia Society website: https://haemophilia.org.uk/

Alder Hey Children’s NHS Foundation Trust: https://alderhey.nhs.uk/

Further booklets and information sheets are available from the Haematology Treatment Room waiting area, with further information from the Haemophilia Society.

Contact Information

Haematology Nurses 8.30am-5.00pm 0151 252 5070 with answer machine, For non-urgent queries which will be reviewed by the end of the next working day.

Nicki Mackett: 07584 234 526 nicki.mackett@nhs.net

Catherine Benfield: 07876 132 163 catherine.benfield@nhs.net

For urgent clinical needs please go directly to Accident and Emergency Department

This leaflet only gives general information. You must always discuss the individual treatment of your child with the appropriate member of staff. Do not rely on this leaflet alone for information about your child’s treatment.

This information can be made available in other languages and formats if requested.

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