Introduction

This leaflet aims to provide you with general information about the signs and symptoms your child may have when they have von Willebrand Disease (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 Type 3 VWD?

Type 3 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 Willebrand 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 VWP protein.
- **Type 2** is found in between 15-30% of people with VWD. This is the reduction in quality of VWP protein. The levels maybe 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 Von Willebrand protein. 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 the most effective treatment for your child. Genetic tests can only be undertaken with consent.

Where did my child’s von Willebrand Disease come from?

Type 3 Von Willebrand disease is inherited from two carrier parents. Both parents have one copy of the faulty gene and may have mild or no symptoms of a bleeding disorder. Rarely a spontaneous genetic error can occur and your child may be the first person in the family known to have type 3 von Willebrand Disease.
Inheritance of von Willebrand Disease

Type 3 VWD is autosomal recessive.

Both parents would have to be carriers of type 3 VWD in order to have a child with Type 3 VWD.

There would be a 25% risk of having a child with Type 3 VWD, 25% of having an unaffected child and a 50% risk of having a child who is a carrier.

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.
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 are the signs and symptoms of type 3 von Willebrand Disease?

- Nosebleeds
- Bleeding gums
- Bruising more than other children
- Prolonged bleeding post dental extractions
- Prolonged bleeding post-surgery
- Bruising or bleeding after vaccinations
- Heavy periods
- Joint and muscle bleeds

**Children with type 3 VWD** can have problems with nose bleeds and bleeding from the gums. Some may have bleeding into muscles and joints. If these are not managed well they can be painful and eventually lead to long term damage.

Traumatic Bleeds: are those which happen after a known injury.

Spontaneous Bleeds: develop when there has been no known injury. Bleeding episodes often need to be treated with replacement VWF, so that the bleeding can be switched off.
How should you ask for advice?

If you are worried about your child please 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 3 VWD.

Situations when you should ask for advice

If your child

- has a significant head injury:
  - for babies: any signs of high pitched crying increased sleepiness decreased alertness, reduced feeding or vomiting.
  - 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
- is showing signs of a joint bleed: Restricted movement in the affected area, swelling, tenderness and heat when touched and pain on movement
- has a heavy or difficult to manage ncontrollable menstrual 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 ensure 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 3 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.

Joint and muscle bleeds. Use normal first aid measures first; this includes Protect Rest Ice Compression Elevation otherwise known as PRICE

Protect and (pain killers) - Make sure your son 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 you 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 a knee, ankle, 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.

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

**VW Factor (VWF)** is given as an injection into the vein. It may be used to stop mouth or nosebleeds or bleeding in to muscles or joints, to manage menstrual periods or during surgery or dental work. It works by replacing the von Willebrand factor in the blood stream bringing the factor levels up to normal so that the bleeding will stop.

**What is Prophylaxis?**

Most children with VWF and Factor 8 levels of 3% or less have a prophylaxis treatment schedule. Prophylaxis is preventative treatment which is given to reduce the risk of minor bleeding into joints and muscles. Most prophylaxis programmes are for treatment 1-2 times a week. The aim of the programme is for the Factor levels to be kept above base level for most of the time reducing the risk of bleeding episodes.

Usually prophylaxis is gradually introduced often starting with one treatment per week increasing to twice per week if required.

There are different types of prophylaxis

- **Primary prophylaxis**: Factor replacement given regularly 1-2 times a week. It is used as long term management of type 3 VWD and is usually started after the first joint or muscle bleed or by the age of two years.
- **Secondary Prophylaxis**: Factor replacement given regularly 1-2 times a week. It is used as long term management of haemophilia and is started after two or more joint bleeds have occurred in order to prevent joint damage.
- **Prophylaxis for activity**: Treatments prior to activities for example every Wednesday before football.

Prophylaxis aims to keep the VWF levels above 3%. When the Factor levels fall below this there is an increased risk of a breakthrough bleed.

**Why give prophylaxis?**

We need to keep your child’s joints and muscles healthy, to do this we need to be able to keep bleeds to a minimum. Recurrent bleeding into a joint causes damage to the soft spongy tissue, the cartilage, and the thin layer of tissue lining the inside of the joint (synovium). Joint damage is more commonly noticed in adults but without a good treatment and prophylaxis plan minor damage can begin at a much earlier age.

**What is a breakthrough bleed?**

Prophylaxis aims to keep Factor levels between 50% (just after the injection) and 3% (just before the next dose of treatment is due). Breakthrough bleeds happen “Spontaneously” despite prophylaxis; they are most likely to occur as the factor level falls to 3% or below.
What is a target joint?

A target joint is one in which 3 spontaneous bleeds have occurred in a six month period. In this situation we would urgently want to evaluate and update prophylaxis and treatment plans.

Treating your child at home

We want you have the opportunity to become independent, but not isolated from Alder Hey. As you become more familiar with type 3 VWD, the signs and symptoms of a bleed and bleed management you may also begin to feel that giving treatment at home is a possibility.

Home Treatment can be helpful for your child, you and the rest of the family this can be helpful for you and the family as:

- Bleeds can be treated more quickly
- You rely less on hospital based treatment giving you more time to be at home, school or work.
- It increases confidence in managing VWD
- You can be more independent

How is treatment given: Treatment is given one of two ways either directly into the vein with a butterfly needle or into a device called a port a cath, ask the nurse specialists for the leaflet “my child needs a port a cath” for further information.

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 he 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 son 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. 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/](https://haemophilia.org.uk/)

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

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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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