

Irish Haemophilia Society



Information for Teachers & Playgroup Leaders



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The Irish Haemophilia Society was founded in 1968 by members of the medical profession, people with haemophilia, their families and friends who felt the need to provide support and advice for members and to improve the quality of life for people with haemophilia.

A representative from the Society is available to present information talks on related topics to schools on request.

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WHAT IS HAEMOPHILIA?

Haemophilia is a lifelong bleeding disorder which slows the clotting mechanism in the blood. It is caused by a deficiency or absence of one of the clotting factors which are necessary to make blood clot.

In Ireland there are approximately 450 males who have haemophilia. Haemophilia in females is very rare.



A person with haemophilia does not bleed any faster than usual. However, it will take longer for the bleeding to stop.

The condition can be mild, moderate or severe and the severity of the condition is directly related to the severity of the deficiency. Factor VIII (A), Factor IX (B) and von Willebrand's factor are the clotting factors most commonly affected.

People with mild and moderate bleeding disorders will have few problems. Their blood clots less efficiently than normal, however this only causes trouble during surgery or after major trauma. People with severe bleeding disorders have inefficient blood clotting and this can cause bleeding even when the person is apparently not injured. Most of these guidelines will apply mainly to people with severe forms of a bleeding disorder.

With appropriate treatment haemophilia can be managed effectively. The most common treatment is an injection of the missing clotting factor into the child's veins. Early treatment is the key to quick rehabilitation.

Some parents have been trained to give treatment to their children. Other children will need to be taken to the hospital for treatment. Bleeding can be internal or external.

EXTERNAL BLEEDING

As per School and Playgroup guidelines, when dealing with open bleeding with any child, you should wear gloves. All blood spillages should be cleaned up using a bleach solution and paper towels which can be disposed of in a strong polythene bag. The top should be securely tied and the whole bag and contents placed into a biological bin (Yellow). The usual first aid measures are usually enough to manage external bleeding.



Cuts and grazes

Cover them with a plaster and bandage and apply pressure to them for a few minutes. Deep cuts that may need stitching will need treatment at a haemophilia centre.

Nosebleeds

Tilt head forward and pinch the bridge of the nose below the bone for 10-20 minutes and/or put an ice-pack on the bridge of the nose for not more than 5 minutes.

Bruises

Children with haemophilia bruise more easily than children without haemophilia and their bruises will be bigger. Bruises only need treatment if they are very painful.

Mouth and Tongue Bleeds

These can be hard to control because clots that form are washed away by saliva, or knocked off by the tongue or food. These bleeds usually need treatment by parents or the treatment centre but try giving the child an ice cube or ice pop to suck as this may do the trick.

If the measures above do not work please contact the child's parents and/or seek medical advice. The child may need coagulation factor concentrate. Be sure to tell the doctor that the child has haemophilia.

INTERNAL BLEEDING

The most common types of internal bleeding are into the joints or muscles and may occur without any obvious cause. Repeated or untreated bleeding into joints and muscles can cause permanent damage such as arthritis, chronic pain and joint damage requiring surgery. Older children should tell you if they have a bleed. Younger children may seem upset, or may protect a limb by limping or not using it.

Signs of bleeding into the joints or muscles are:

- The child will be reluctant to move the affected limb/joint.
- The child will complain of pain in the affected joint/limb.
- There may be swelling and heat in the affected joint or limb if the bleed is not treated promptly.



Treatment for bleeding into the joints and muscles is:

Coagulation factor concentrate (IV), **R**est, **I**ce, **C**ompression and **E**levation (**C-RICE**).

Joint/muscle bleeds should be treated as soon as possible. If a child complains of any of the above signs please contact the child's parents and/or seek medical advice, elevate the limb and apply ice (if available) and do not allow the child to use the limb until his parent/guardian arrives. Do not apply pressure until the child has had coagulation factor concentrate as constricting swelling can be dangerous.

Children can sometimes return to school when recovering from a bleed. However, they should be allowed to elevate the affected limb and be excused from activities such as PE.

RARER BLEEDING EPISODES

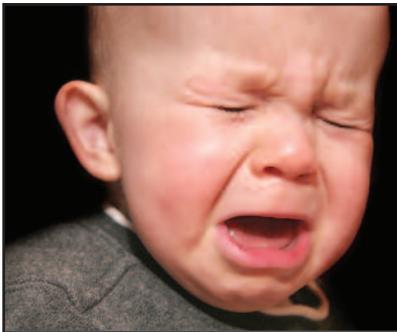
Bleeding in the gastrointestinal (GI) tract (gut) is very unusual in haemophilia. If a child cough's or vomits up blood or reports blood in his stool or urine you should contact the child's parents and/or seek medical advise.

Head, face, neck and throat, injuries may be potentially life threatening (if vital organs are affected). Contact the child's parents and/or seek medical advise. All head injuries should be treated with coagulation factor concentrate without delay.

There are 3 sorts of head injuries which should be treated in different ways.

1. Bumps on the Head

Young children bump their heads. These bumps do not cause bleeds. If the child is not upset by the bump and shows no signs of pain, the injury is unlikely to be serious and probably does not need treatment. It may be difficult to decide whether a head bump is serious or not. If in doubt, get treatment.



2. Minor Head Injuries

These are injuries that lead to bruising or even small cuts on the child's head. These injuries should always be treated, either by the parent or the haemophilia centre.

Either of the above can become a serious head injury. Therefore, a careful eye should be kept on children for at least 12 hours for any of the symptoms listed on the next page.

3. Serious head injuries

These result from a severe bump on the head. A head injury is always serious if the person is knocked unconscious. These injuries should be treated as quickly as possible and the child should be taken to the nearest haemophilia centre.



Signs of a serious head injury

If the child has any of these signs, regardless of the apparent injury, they may have a serious head injury. Some signs include:

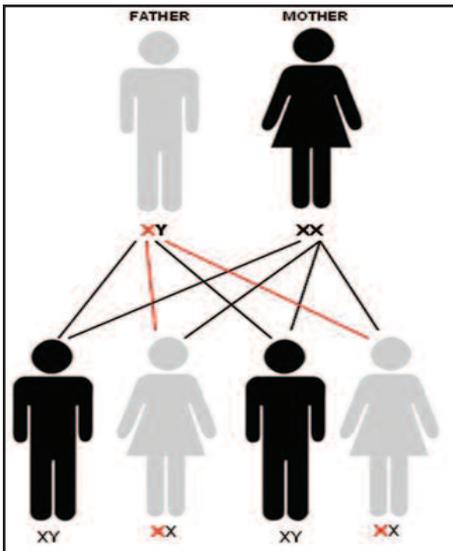
- A persistent or increasing headache.
- Vomiting.
- Sleepiness or a change in his normal behaviour.
- Weakness or clumsiness in legs or arms.
- Stiffness or pain in the neck.
- Blurred or double vision, or going cross-eyed.
- Poor balance.
- Seizures or convulsions.

In the event of an emergency phone the haemophilia treatment centre and parent immediately.

INHERITANCE OF HAEMOPHILIA AND VON WILLEBRAND'S DISEASE

Haemophilia is an inherited condition. The gene responsible for Factor VIII and Factor IX production is carried on the X chromosome. Females have two FVIII and FIX genes and men have one. Therefore, men are affected and women are carriers. The diagrams below may assist in understanding this.

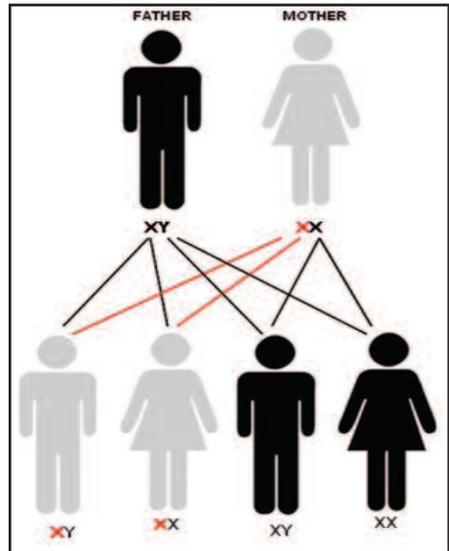
When the father has haemophilia and the mother is unaffected



None of the sons will have haemophilia.

All of the daughters will carry the haemophilia gene.

When the mother carries the haemophilia gene and the father is unaffected



There is 50% chance at each birth that a son will have haemophilia.

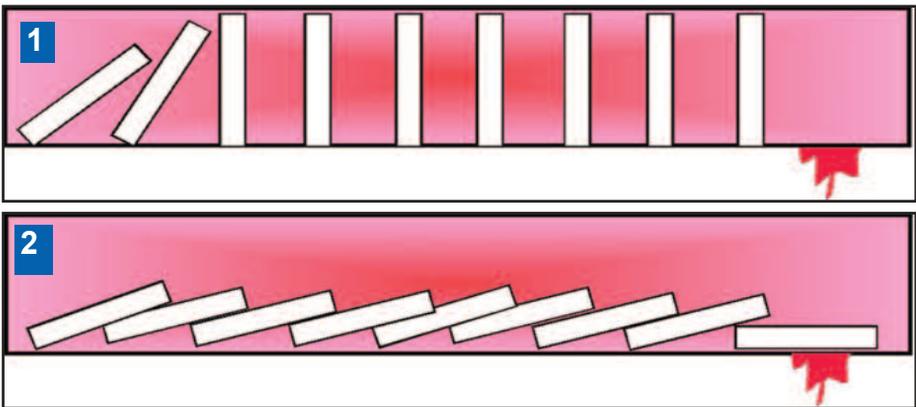
There is a 50% chance at each birth that a daughter will carry the haemophilia gene.

Von Willebrand's is slightly different. Both men and women have two von Willebrand factor genes. Men and women are equally affected by von Willebrand's disease (vWD). VWD is usually mild although it can be severe. Most people with vWD have the Type 1 or Type 2 form and will only experience a mild form of the condition. No medication will be required except when having surgery, or dental extractions.

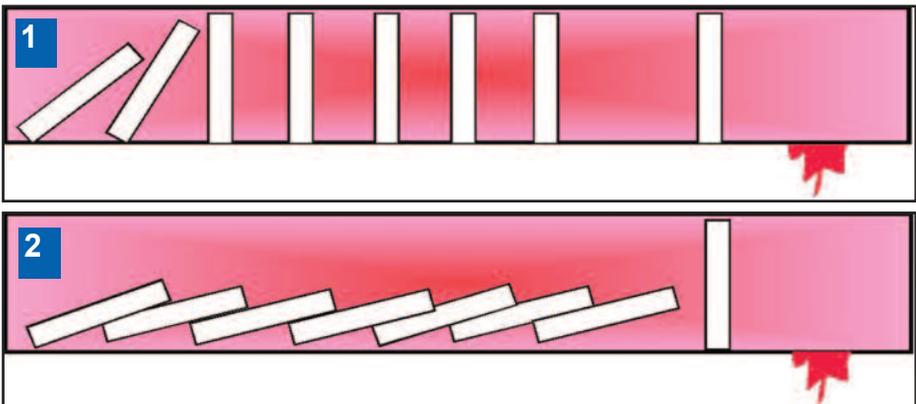
THE CLOTTING MECHANISM

Bleeding is controlled by a number of clotting factors. There are 12 clotting factors that act together in a “domino effect” to clot the blood and repair any damaged blood vessels. In haemophilia, vWD or other bleeding disorders this mechanism does not work. When a person with haemophilia or vWD has a cut, the first stages work normally to repair the damage and when it reaches the stage where the person is deficient in a specific factor, the “domino effect” is stopped and the clot is not formed. So it takes longer for the bleeding to stop.

NORMAL CLOTTING



HAEMOPHILIA





EDUCATION

Academically there is no difference between a child with a bleeding disorder and an unaffected child. Haemophilia, von Willebrand's or a related bleeding disorder should not interfere with a child's education. Most severely affected children receive preventative treatment 2-3 times weekly at home which prevents most bleeding episodes. This is known as prophylaxis and requires intravenous (i.v.) injections given by the parents or the child themselves. It is important for them to attend school as much as possible. However, sometimes they may miss school when they are recovering from a bleed. Teachers should do their best to make them comfortable when they come back, and to help them catch up on work they have missed.

Another important aspect is the development of a 'trusting' relationship between teacher and child as if the need for treatment should a bleed occur during school time. If the child continually claims to have a bleed during or before a particular lesson, the teacher would be suspicious that it is genuine.

All these issues should be discussed at initial meetings with parents so that provisions can be made to deal with any difficulties that may arise.



PLAYGROUP AND PRIMARY SCHOOL

At playgroup, normal play activities present little in the way of problems and the child should be allowed to play alongside the other children.

Some unsightly bruises may occur, but in general these will not be a serious problem and no treatment will be necessary. There is no reason why children with a bleeding disorder should not be allowed to use scissors and other sharp instruments. Like any child, they need to learn how to handle this equipment.



SECONDARY SCHOOL

Primary school level sport is usually less competitive and children with haemophilia and von Willebrands should be allowed to take part in all activities unless they have specific problems.

In secondary school sport often becomes more competitive and injuries are more common. Sports with a high level of physical contact such as rugby or boxing are not recommended. Sports booklets are available for more details.



SPORTS AND LEISURE

Appropriate sports and physical activities are generally recommended. Almost all sports and leisure activities are safe. Children with haemophilia and bleeding disorders should play sports, just like their friends do. Exercise will promote strong joints and muscles so they are less likely to bleed.

It is important to encourage people with haemophilia or von Willebrand's to explore new physical challenges gradually and not to dash into potentially harmful activities. Only time will tell, if a particular activity is going to provoke unacceptable bleeding, and the chances of this happening will be reduced by careful attention to warm-ups, stretching exercises and cool-downs.

If it is found that a particular sport causes bleeds, it may be necessary to change to another sport or take treatment before taking part.

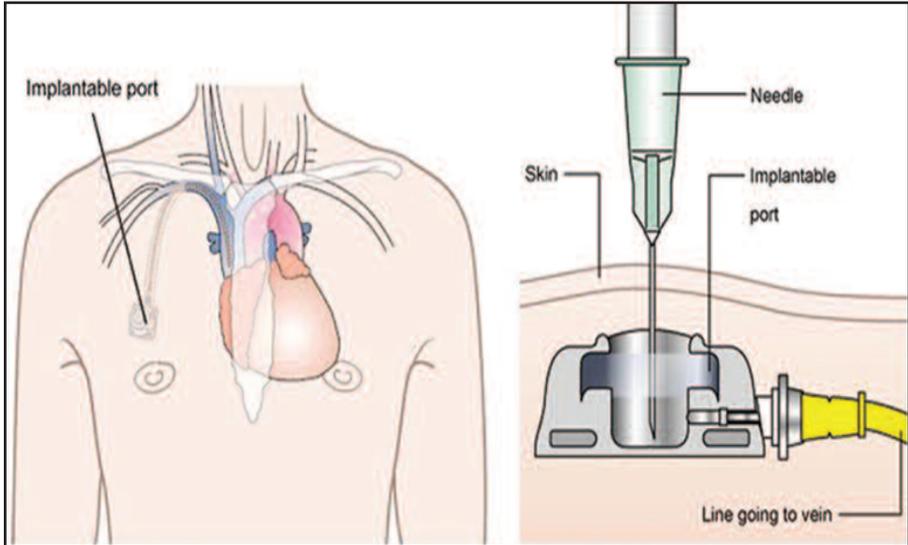
Sports injuries occur just as much to people with a bleeding disorder as to anyone else. The treatment is the same.

When a bleed occurs the rules governing treatment are very simple:

- The earlier a bleed is treated the better.
- If in doubt, treat.

PORT-A-CATH

Some children do not have good veins and may have a Port-a-Cath to facilitate their parents giving factor at home. This device is a small bung under the skin which connects directly into the blood stream. They can be placed in the chest or arm. There will be a small scar at the site of insertion and you may see a small bulge under the child's skin. Injections are made by pushing a special needle through the skin into the port.



Some basics to remember

People with Haemophilia or vWD:

- Bleed no faster than normal.
- Are in no danger from pinpricks, scratches or grazes (because their first line of defence against bleeding is still intact).
- Usually bleed internally into joints or muscles.
- May recognise when a bleed has started, often long before there are any signs (such as swelling).
- Respond rapidly to clotting factor replacement.
- Have a normal physique, unless they have had inadequate treatment which has resulted in arthritis. This is caused by repeated bleed into a joint.

FREQUENTLY ASKED QUESTIONS

Are there different types of haemophilia and von Willebrand's?

Haemophilia can be mild, moderate or severe, depending on the degree of the deficiency (i.e. how much factor is missing). There are 3 types of von Willebrand's disease and these are classified as Type 1, Type 2 and Type 3. Type 3 is the most severe however the vast majority of cases are either Type 1 or Type 2 and are rarely severely affected.

Will anyone who has haemophilia or von Willebrand's bleed profusely from any cut or injury they sustain?

People with haemophilia, von Willebrand's disease or a related bleeding disorder will not bleed any faster than anyone else with a similar injury. Because the clotting mechanism is not intact they will bleed for longer than normal and may require treatment for the injury.

Should children with a bleeding disorder be allowed to use scissors or sharp instruments?

Every child has a need to be taught how to handle and use sharp instruments so that they are aware of the dangers of cutting themselves.

Will reducing activity reduce the risk of injury and bleeds?

The less active a person with a severe bleeding disorder is, the more likely they are to suffer bleeds. Physical activity strengthens bone and muscle. Exercise improves muscle tone and co-ordination. Strong muscles protect vulnerable joints.

Can people with haemophilia or von Willebrand's play sport?

Yes. Exercise is good for everyone. People with haemophilia or von Willebrand's will find that sport strengthens joints and muscles, helping prevent bleeds. Care needs to be taken when choosing a sport, as rough high contact sports such as football and boxing aren't usually recommended as they may cause bleeds. People learn to manage their condition know what is good for them to do and are recommended to speak to their doctor.

Should a child with haemophilia or von Willebrand's wear protective gear, helmets etc?

No. Current treatments mean that protective gear for every-day living is not necessary. Like all people, protective gear/helmets should be worn for activities such as horse riding motorbikes and bicycles, skateboarding and football.

Do people with haemophilia or von Willebrand's die young?

No. Treatments currently available allow people live a normal lifespan.

Is haemophilia or von Willebrand's contagious?

Haemophilia or vWD cannot be transmitted to anyone else. It is a genetically inherited disorder the only way it can be transmitted is through pregnancy (from mother to

PERSONAL INFORMATION FOR TEACHERS & PLAYGROUP LEADERS

Name of Child:	
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Date of Birth:	
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Address:	
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MOTHER

Home Phone:		Mobile:	
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FATHER

Home Phone:		Mobile:	
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EMERGENCY CONTACT

Name:		Contact No.	
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Bleeding Disorder:	
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Condition is:	Mild <input type="checkbox"/>	Moderate <input type="checkbox"/>	Severe <input type="checkbox"/>
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Normal Treatment:	
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Haemophilia Treatment Centre:	
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Name of Doctor / Consultant / Nurse:	
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Contact Numbers:	
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