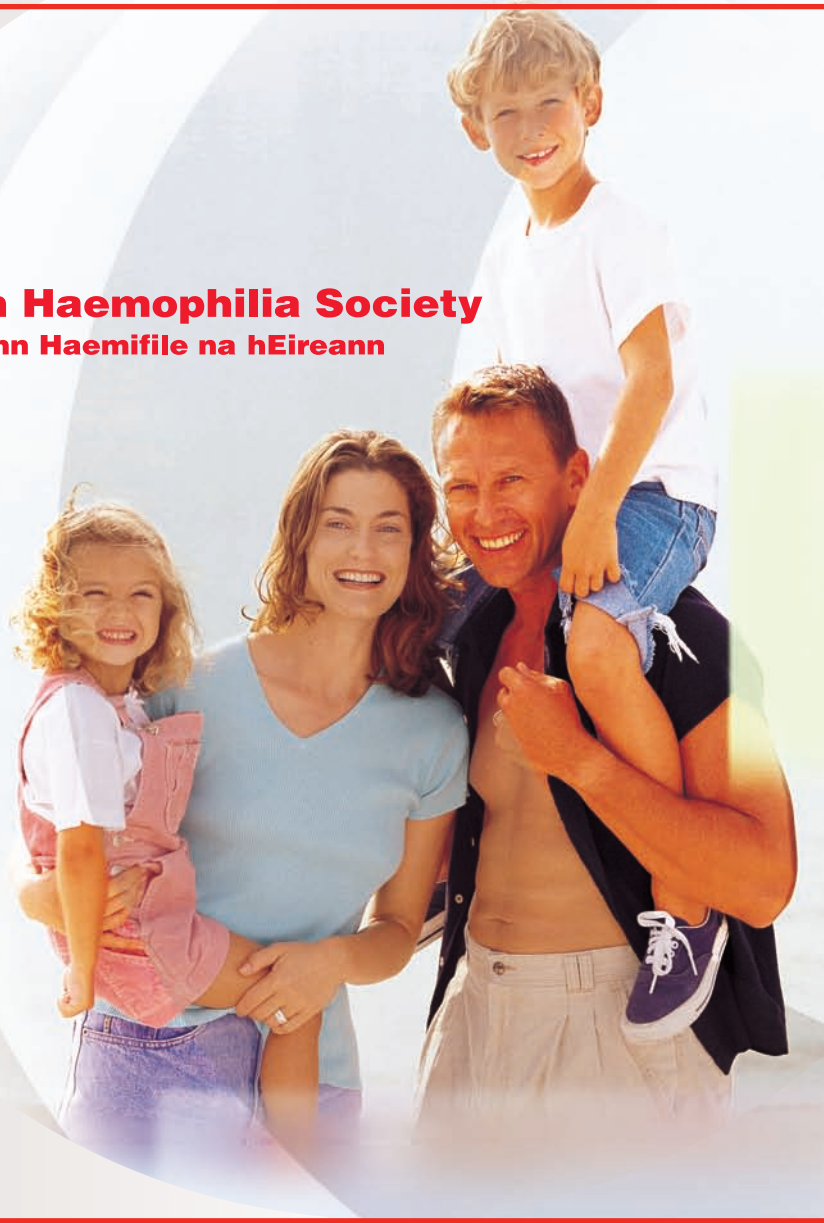
The Irish Haemophilia Society
Cumann Haemifile na hEireann



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Introduction to Haemophilia & Related Bleeding Disorders

Compiled and edited by
The Irish Haemophilia Society

USEFUL CONTACTS

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CONTENTS

WHAT IS HAEMOPHILIA?

The general term "haemophilia" describes a group of inherited blood disorders in which there is a life long defect in the clotting mechanism of the blood. Haemophilia is quite rare. The incidence of haemophilia in the general population is 1 in 10,000 (therefore about 1 in 5,000 of the male population has haemophilia)

A more precise definition can be given in terms of which part of the clotting mechanism is defective. The clotting factors, which are present in blood are numbered with Roman Numerals from I to XIII.
In normal blood the clotting factors act like a row of dominoes toppling against each other to create a chain reaction (figure 1)

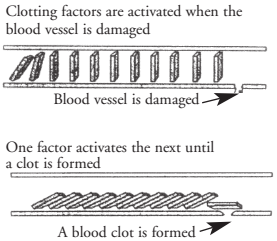


Figure 1

When one of the factors is missing the clotting mechanism is not activated (figure 2)

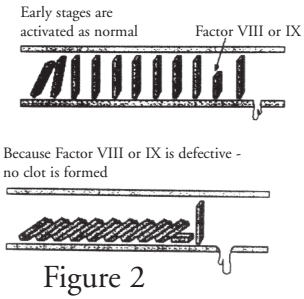


Figure 2

A person can be deficient in any factor but most commonly in Factor VIII and Factor IX.
A person with Haemophilia A is deficient in Factor VIII.
A person with Haemophilia B (or Christmas Disease) is deficient in Factor IX.

Both types of haemophilia share the same symptoms and inheritance pattern, only blood tests can differentiate which factor is affected. The severity of the condition is related to the degree of deficiency of the relevant clotting factor in the blood.

There are other related bleeding disorders where the person is deficient in other clotting factors. (see table page 10)

Severity

The severity of the haemophilia condition is related to the degree of deficiency of the relevant clotting factor(s) in the blood. Normal clotting factor activity is described as between 50% and 200%.

Mother carrying hemophilia gene:
A person with less than 1% of normal clotting activity is described as "severe".
A person with between 1% and 5% of normal activity is described as "moderate.
A person with over 5% but less than 50% of normal activity is described as "mild".

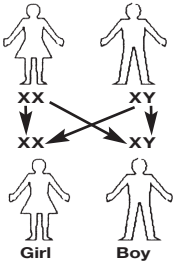
Those who have mild or moderate haemophilia generally only experience bleeding problems after an obvious injury or an operation, and many mild cases only experience bleeding after, for example, a tooth extraction or surgery.

INHERITANCE PATTERN

Haemophilia is an inherited condition. However, it is possible for the condition to appear in any family – it is thought that at least 30% of people with haemophilia have no family history of a bleeding disorder. It is difficult to be exact about this because of the way in which haemophilia is inherited. Technically, it has a "sex linked recessive" inheritance pattern.
This means that while only males are affected by the condition, it is passed through the female member of the family.

The sex of a newly conceived baby is determined by the type of chromosomes it receives – one from each parent. A boy inherits his mother's X chromosome and his father's Y chromosome, and a girl has two X chromosomes, one from each parent (see figure 3)

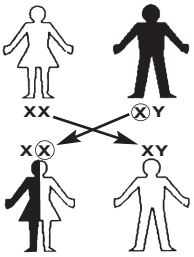
The defect that causes haemophilia rests in the X chromosome, therefore, all daughters of a man with haemophilia will inherit his X chromosome and will be a carriers of the haemophilia gene.



(figure 3)

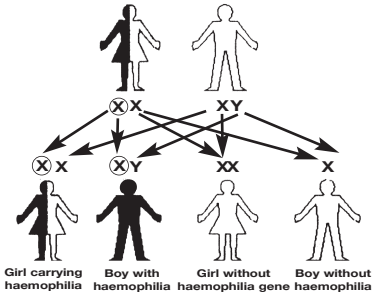
Since the sons of a man with haemophilia must inherit his Y chromosome they will not be affected by haemophilia nor will they be carriers.

A carrier female has one normal and one defective "X" chromosome. If she has a son, the son has a 50:50 chance of receiving his mother's defective "X" chromosome. (see figure 4)



(figure 4)

The daughter of a carrier also has a 50:50 chance of being a carrier herself.
(see figure 5)



(figure 5)

Although the daughter of a man with haemophilia will not have as low levels as her father, she may still have a lower than "normal" level.
Because of the ability for the defective gene to remain hidden through several generations of carrier daughters it can be difficult to know whether there really is no family history, or whether the problem has just been dormant for several generations.
(see section on Carrier Detection page 15)

Diagnosis

A blood sample from the newborn baby can be used to make a diagnosis. This applies to mild, moderate and severe cases. In at least 30% of cases of haemophilia there is no known family history and the occurrence of haemophilia is presumed to be the result of a spontaneous genetic mutation.

Cases of severe haemophilia may become apparent and be diagnosed at an early age as a result of surgery or injury. For example, prolonged bleedings may follow circumcision, routine blood sampling or routine childhood vaccinations.

More often the first symptom of a bleeding tendency is in the form of extensive bruising as the child learns to crawl or walk. Unfortunately this is sometimes suspected to be a result of non accidental injury, but increasingly in such cases it is automatic that coagulation tests are used to investigate the possibility of a child having a bleeding disorder. Cutting teeth is another stage at which the condition may become apparent.

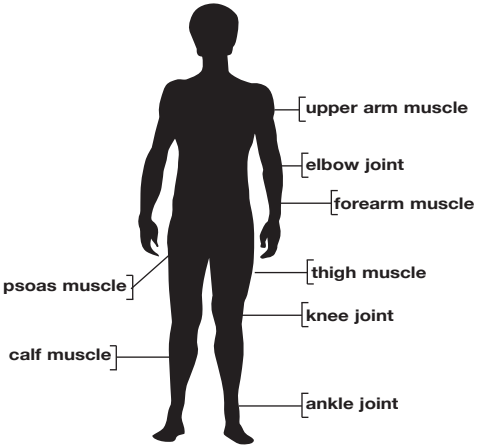
Moderate and mild haemophilia may not be diagnosed until later in childhood or in some cases even in adulthood. Because there is some clotting factor available more minor injuries will heal normally and it may not be until a major injury occurs that the deficiency is revealed. The process of diagnosis involves many complex laboratory tests on blood samples and takes several days to complete. The time around diagnosis can be a difficult period for parents particularly where there is no family history of the condition. It is important that parents become informed about haemophilia and the impact it is likely to have on their child and other members of the family. The Irish Haemophilia Society is available to offer support to all family members and provides a range of education and support programmes for all age groups.

Signs & Symptoms of bleeding

Cuts and Scratches: In most cases minor cuts and scratches do not pose any problems for a person with haemophilia. A little pressure is usually enough to stop the bleeding. A person with a bleeding disorder does not cut more easily, or bleed more profusely or bleed faster than normal.
They simply bleed for longer.

Joints and Muscles: For those severely affected, the major problem is internal bleeding into joints, muscles and soft tissues. All of us damage our muscles in small ways in the activities of everyday life. Most people repair that damage automatically and unnoticed. For the person with a severe bleeding disorder, however, the tiny breaks in the blood vessels in joints and muscles may continue to bleed as a result of trivial twists and strains. These bleeds are sometimes described as “spontaneous” because it is impossible to identify the exact injury which caused them.

Bleeding occurs most often inside these joints or muscles.



Soft Tissue Bruises: An ache, irritation or tingling in an affected area is the usual indication that a person with haemophilia gets when bleeding into the body tissue begins. Then follows definite pain and stiffness, limitation of use, and the site of the bleed, will get hot, swollen and progressively more tender. If left untreated the pain becomes excruciating. In the case of joint bleeding, the blood which has escaped into the joint has a very damaging effect on the surface of that joint. Once a joint becomes damaged then bleeding will occur more frequently resulting in a "target joint". The majority of bleeds into joints and muscles occur in the lower limbs, with knees and ankles being the worst affected in most people. Soft tissue bruises will always occur in people with bleeding disorders. Although these may look serious they usually do not require any treatment. Sometimes if the bruise is increasing in size and causing pain, then factor treatment may be recommended.

Head, Face and Neck Injuries: After a blow to the head, face or neck sufficient factor must be given immediately to bring the factor level to 100%. All these injuries must be assessed at a treatment centre. If the injury is significant enough a person with a bleeding disorder may need to be admitted to hospital and may require a CT scan to ascertain the extent of bleeding, if any.

Minor Head Injuries: These are injuries that can lead to bruising or even small cuts on the head. These injuries should always be treated either at home or at the treatment centre.

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 patient taken to the nearest haemophilia centre.

Other Serious Bleeding Episodes:
Vomiting blood
Coughing up blood
Passing blood in bowel motions (stools will be black in colour)

TREATMENT

Each bleeding episode must be promptly treated by an intravenous infusion of the clotting factor which is deficient. Once the bleeding stops, pain rapidly diminishes and use of the limb returns. The concentrate is administered as an intravenous injection, which is manufactured as a white powder and should be reconstituted with the sterile water, provided with the factor.

The Factor VIII and Factor IX used for the treatment of haemophilia is called "recombinant factor". Recombinant Factor VIII and Factor IX products used in Ireland are produced by cell lines using re-combined DNA.

What cannot be emphasized enough is that a person with haemophilia must have treatment as soon as a bleed starts. It prevents further bleeding and pain and, most importantly, reduces the likelihood of permanent damage to joints (target joints).

Surgery

All surgical procedures, including dental extractions and fillings, will require treatment beforehand and possibly for 1-6 days afterwards. This should be organised with the Treatment Centre.

Home Treatment

Home treatment is the ideal method of treatment from a medical viewpoint as a minimum amount of time is lost between the recognition of a bleed and treatment. This has many advantages. It reduces the disruption caused by a bleeding episode to the person with haemophilia and his family and the patient feels more able to control his condition.

The benefits of home treatment not only include increased independence and the bonus of not having to travel to the hospital at all hours for treatment, but school and work attendance is more regular and participation in social activities is easier. If bleeds are treated promptly the period of incapacity caused by each episode can be reduced. In adults and teenagers, this procedure is usually carried out by the affected person. In younger children this will be taught to parents and in babies a device called a Port-a-cath can be used to facilitate venous access.

Prophylaxis

It is recommended that all children with severe factor deficiencies should be commenced on a programme of factor "prophylaxis" before their second birthday or earlier if they have experienced any joint bleeds. Prophylaxis involves small regular (2-3 per week) injections of factor to prevent spontaneous bleeding and to minimize traumatic bleeding. This treatment regime, although time consuming and sometimes difficult to learn, will prevent joint damage and lead to an improved quality of life.

Port-a-Caths

In some young children where venous access is difficult and injections are stressful for both the child and the parent it is recommended that a Port-a-Cath be fitted. Port-a-Caths are used to provide access to veins and are inserted under general anaesthetic. The Port-a-Cath is inserted under the skin usually in the chest area. When the Port has been inserted there is no longer a need for injections into the veins so treatment is painless and therefore less stressful.

Inhibitors to factor concentrate

Some people with severe haemophilia develop an antibody or 'inhibitor' to factor treatment. The effect of this is that when the factor is given for a bleed there is no clinical response because the activity is immediately neutralised. Often the first sign that a person has inhibitors is that his treatment does not appear to be working.

People who have inhibitors to Factor VIII or Factor IX have to be given treatment with a recombinant factor called Factor VIIa (Novoseven). This is an activated factor, which bypasses the need for Factor VIII or Factor IX in the clotting cascade. It is given by intravenous injection but only gives very short-term response, therefore needs to be given every 2-3 hours. Eradicating Factor VIII or Factor IX inhibitors involves highly specialised and intensive treatment regimes, which may not always be successful. It is good practice to screen small children with haemophilia regularly for inhibitors.

Pain Management

It is important to treat bleeding episodes without delay as this will minimize the pain caused by internal bleeding. Before prompt and adequate treatment became available in the 1970's pain was a regular feature of life for people with severe haemophilia. Untreated bleeds resulted in excruciating pain and arthritic deterioration. In the long term arthritic deterioration can lead to constant pain in a damaged joint. Many people with haemophilia still have constant pain as a result of arthritic damage to their joints. You should follow your doctor's advice about effective medication to ease your pain and take your medication as prescribed. If you are experiencing problems with the medication or it is not working for you contact your doctor again. Remember that some medication should be taken at certain times i.e. before or after food. Find out the side effects, if any, of your medication and what to do if they occur. Take your doctors advice before using "over the counter" medications.

Remember: People with bleeding disorders should not take aspirin in any form as this can aggravate bleeding.

Safety Issues

In the past, the blood products used for the treatment of people with haemophilia contained many other ingredients than the Factor VIII or Factor IX that the patient required. This included many contaminants, some of which caused liver damage and disease. One of the most serious was Hepatitis. Hepatitis A was seldom a problem, but the more serious Hepatitis B and Hepatitis C viruses were found in many people with haemophilia. In many cases there were no symptoms but tests revealed that the hepatitis virus was present and liver function was impaired. In Ireland 220 people with bleeding disorders were infected with Hepatitis C. Approximately 20% of people with haemophilia or von Willebrand Disease, who regularly received blood products, had serious liver disease including cirrhosis.

The most serious problem for people with hemophilia was the threat of infection with HIV and the subsequent development of AIDS. The haemophilia community in Ireland was devastated by the infection of 106 men and boys with HIV through injection with contaminated blood products.

Hepatitis and HIV gave rise to many new fears and problems just at a time when life for people with haemophilia appeared to be better than it had been in the days before effective treatment was available. It was essential for people who were infected and their families that counselling and support services were put in place and the Society responded by introducing a range of support programmes to meet the needs of the members.

Currently recombinant factor replacement therapy is used to treat Factor VIII and Factor IX deficiency. In excess of 1 billion units of recombinant factor concentrate has been used worldwide with no reported case of HIV or Hepatitis. All other factor deficiencies (excluding vWD in most cases) are treated with products which are made from human donated plasma. Lessons have been learnt from the past. Nowadays blood products are safer than ever before as they are extensively screened for viruses and treated to inactivate any viruses which may be present. Patients are also offered a vaccination programme against Hepatitis A and B. Vigilance must be maintained.
The Society now has a formal role in the selection and tender process for all treatment products for Haemophilia, von Willebrand Disease and other bleeding disorders. Two representatives of the Irish Haemophilia Society are members of the Product Selection Monitoring and Advisory Group (PSMAG). The PSMAG formally selects the products used for the treatment of Haemophilia, von Willebrand Disease and related bleeding disorders from all the products available. The group also monitors developments in relation to these products on an ongoing basis. The Society has an independent expert advisor who attends meetings of the PSMAG as necessary and who advises the Society in relation to their work on this committee.

Von Willebrand Disease

von Willebrand Disease (vWD) is the most common inherited bleeding disorder and affects both men and women. People with vWD are deficient in von Willebrand factor (vWF). A person with vWD may bleed for longer than normal, although they do not bleed any faster than normal.

The severity of vWD varies, but it usually affects people only mildly. Therefore, most people with vWD live completely normal, active lives. People with vWD usually have some vW factor in their blood, just lower levels than normal.

In these people the disease is mild and the common symptoms are easy bruising, frequent and sometimes severe nosebleeds and unusual bleeding from the mouth or gums. Gastrointestinal or urinary tract bleeding may also be a problem. In women vWD can cause heavy periods. (see section on women with bleeding disorders)

Types of von Willebrand Disease

Type 1 – This is the mildest and most common form of vWD. Those diagnosed with Type 1 vWD have low levels of von Willebrand Factor.

Type 2 – In type 2 vWD the von Willebrand Factor does not work properly. Type 2 is divided into type 2a, 2b, 2m and 2n. These types are treated in different ways. Some people with type 2 can also have low amounts of von Willebrand Factor.

Type 3 – This is the most severe and also the rarest form of vWD. A person with type 3 usually has little or no detectable level of vWF and usually has low amounts of clotting factor VIII. This can cause symptoms like those of haemophilia such as bleeding into the muscles and joints.

Inheritance pattern

vWD is passed on from parents to their children through an abnormal gene. Unlike haemophilia (which is passed on by women and only affects men), vWD affects both women and men, and can be passed on from either the mother or father.

Diagnosis

Because vWD is usually mild, many people do not experience any problems, and the condition may be undiagnosed for a long time. Often, the first indication that there may be a problem with blood clotting comes after a minor operation, such as a tooth extraction.

Once the problem is suspected, doctors can run tests to diagnose the disease. The tests consist of taking a personal and family history of diseases from the patient, which may give clues to clotting problems in the past, or in relatives, and some blood tests. These tests measure the time that blood takes to clot and the levels of clotting factors present, and compares them with those in people known not to have vWD.

The diagnosis may be complicated and take some time because many people with vWD have levels of clotting factors in their blood that are nearly as high as normal. Worrying about going to hospital and having a blood test can also make these levels increase. Therefore, several different tests may be needed to make a final diagnosis and this can take time, and repeat tests are often needed to clarify the diagnosis in mild cases. Diagnosis in one family member can often lead to the identification of the disease in other family members.

Symptoms

Common symptoms are frequent nosebleeds, easy bruising and, in women, heavy periods. Most people with vWD live completely normal, active lives. People with severe forms of vWD can experience symptoms like those in haemophilia such as bleeding into muscles and joints.

Treatment

In mild von Willebrand Disease the body has large 'stores' of Factor VIII and von Willebrand Factor. To release these stores into circulation an IV medication called DDAVP is used. DDAVP is a chemical and is not made from blood products. Administration of DDAVP is by intravenous infusion over approximately 30 minutes. A test dose is always given at diagnosis to document a positive response. DDAVP is also available as a nasal spray, which is recommended for home use.

Sometimes a person does not respond as expected to DDAVP. In this situation plasma derived factor containing Factor VIII and von Willebrand Factor must be used.

RARE BLEEDING DISORDERS

FACTOR	BLEEDING SYMPTOMS	TREATMENT
Factor V		
Mild	Post Trauma / Post Surgery	Fresh frozen Plasma
Moderate	Post Trauma / Post Surgery	Fresh frozen Plasma
Severe	Post Trauma / Post Surgery	Fresh frozen Plasma
Factor VII		
Mild	Mucosal Bleeding, Nose Bleeds, Heavy Periods, Bruising, Bleeding post surgery	Recombinant Factor VII
Moderate	Mucosal Bleeding, Nose Bleeds, Heavy Periods, Bruising, Bleeding post surgery	Recombinant Factor VII
Severe	Mucosal Bleeding, Nose Bleeds, Heavy Periods, Bruising, Bleeding post surgery	Recombinant Factor VII
Factor X		
Mild	Bleeding after trauma or post surgery	Plasma Derived Prothrombin Concentrate
Moderate	Bleeding after trauma or post surgery	Plasma Derived Prothrombin Concentrate
Severe	Spontaneous bleeding from birth Joint/Muscle bleeds	Plasma Derived Prothrombin Concentrate
Factor XI		
Mild	Bleeding after trauma	Fresh frozen Plasma
Moderate	Bleeding after trauma	Fresh frozen Plasma
Severe	Bleeding after trauma	Fresh frozen Plasma
Factor XII	No bleeding associated with this deficiency	No treatment required

The number of people in Ireland currently diagnosed (October 2004) with bleeding disorders is shown in the table below

Bleeding Disorder	Severity	Total
Haemophlia A		
Factor VIII	Severe	156
	Moderate	17
	Mild	179
Haemophilia B		
Factor IX	Severe	45
	Moderate	21
	Mild	118
Von Willebrands		
Disease		540
Factor I		23
Factor V		33
Factor VII		32
Factor X		45
Factor XI		36
Factor XII		125
Factor XIII		3

Source: National Centre for Hereditary Coagulation Disorders, St. James's Hospital, Dublin.

YOUR CHILD WITH A BLEEDING DISORDER

When a child is diagnosed with a bleeding disorder, the parents may have feelings of disappointment, worry and perhaps even guilt. These emotions are temporary. This may be an especially hard time for those who have no family history of the condition. Some people talk about things while others 'bottle up' their worries. Parents and other members of the extended family need to have the reality of haemophilia carefully, patiently, and repeatedly explained to them by members of the comprehensive haemophilia care team. After a while, parents may even find that it is not nearly as bad as they first imagined…the unknown is often more frightening than reality.
The impact of replacement therapy on the lives of people with haemophilia needs to be explained. Likewise when a Port-a-Cath is recommended because of difficulty with venous access, the surgical procedure needs to be fully explained to the parents.

It is important to remember that with good treatment the boy with haemophilia has every chance of growing up as an active, fit child who can participate in family, school and later working life.

Because bleeding disorders are rare conditions parents may feel isolated and alone and it is very helpful to be put in touch with others in a similar position.

The early days

A big worry for parents of a baby with a bleeding disorder is how they can recognize when their child has a bleed.
An infant or young child with a bleed may appear to be irritable for no apparent reason.
He may avoid using the affected limb.
The area may feel warm or swollen.
His limbs may look uneven.
If in doubt you should call your haemophilia centre for advice.
With experience and a little vigilance parents will soon learn to recognize the signs and severity of bleeding episodes and learn to respond effectively.

Developing Social Skills

It is important for the child and for the other members of the family that haemophilia does not dominate the life of the family. A child with haemophilia has to learn to become independent in the same way as any other child. While it is natural for parents to want to offer as much protection as possible, over protection is not in anyone's interest.

Obviously at the toddler stage, when all children are liable to cut, bump and bruise themselves, careful observation of the child is important. However, as children get older they can become more involved in self care and the recognition of bleeds. Young children with bleeding disorders very quickly learn to recognize when a bleeding episode is starting in a joint or muscle. They should be encouraged to tell a parent as soon as possible so that treatment can be given and disruption to normal life reduced to a minimum.

Education

School will present the child and his parents with a new set of challenges. It is important that boys with haemophilia learn social skills in the same manner as other children.

Playgroups and pre school groups can provide a useful introduction to mixing with other children away from the home environment. At playschool normal play activities present little in the way of problems and the child with haemophilia should be allowed to play alongside the other children.

Children with bleeding disorders attend, and cope very well in ordinary schools. Education is vitally important in today's world as it provides the child with every opportunity to participate fully in activities of normal daily life and work.

Haemophilia is rare, so the school staff may not be familiar with the condition. It is vitally important that the head teacher is fully informed of the nature of haemophilia and has an opportunity to discuss the individual boy's condition with his parents. It is not necessary for a child with a bleeding disorder to be singled out for special attention at school.

Sports

Appropriate sports and physical activities are generally recommended for people with haemophilia. It is important to develop strong joints and muscles. It is important to encourage people with haemophila to explore new physical challenges gradually, and not to dash into potentially harmful activity. 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.

Primary school level sport is usually less competitive and boys with haemophilia should be allowed to take part in all activities unless they have specific problems such as a target joint. A target joint is one which is particularly prone to bleeds.

Secondary school sports and activities are almost always safe for boys with haemophilia. They should play sports just like their friends do. The exercise will help make joints and muscles strong, so they are less likely to bleed. If it is found that a particular sport causes bleeds, it may be necessary to change to another sport or take factor replacement therapy before taking part.

However, highly competitive contact sports such as rugby and hurling should be avoided at both primary and secondary level.

College/Employment

On a programme of prophylaxis, a child with haemophilia should not miss out on schooling or education as a result of bleeds or joint damage. The choice of college education and employment, therefore, for someone with a bleeding disorder should not differ from any of his peers.

In each case, the choice of occupation is related firstly to the interests, abilities and skills of the individual and secondly to the severity of haemophilia.

It must be emphasized that the majority of men with haemophilia find and keep satisfactory regular employment. There are often other difficulties unrelated to haemophilia to overcome when an individual is unable to find or keep work.

It is important that prospective employers have the opportunity of understanding what haemophilia means. The vast majority of doubts and prejudices, as far as the employer is concerned, stem from ignorance as to the nature of the condition.

Home treatment has enabled the person with haemophilia to self infuse concentrates at his place of employment, which ensures normal work attendance and promotes self confidence with independence.

Some Points to Remember

You may suspect your child has a bleed but you are not sure. If in doubt assume it is a bleed and treat it as such.

All bumps or knocks to the head should be treated seriously. You should bring your child to your treatment centre immediately – do not wait for symptoms to appear.
Do remember that any medication containing Aspirin should not be given to anyone with a bleeding disorder as this can cause bleeding. For pain relief you should try paracetemol following instructions for the correct safe dose.
Other drugs can affect clotting too. Always check with your doctor which medicine is safe.

Avoid injections into muscles. A muscle injection could cause painful bleeding. However, vaccinations are important and safe for a person with haemophilia. If in doubt contact your Haemophilia Treatment Centre for advice.

When planning your family holiday remember to make contact with your haemophilia centre to arrange to get sufficient factor concentrate for the duration of your stay. If you are travelling outside the country you will need, for customs purposes, to carry a letter from your child's haematologist concerning the product you are carrying.

It is also wise to find out before you travel the location and contact details of the nearest haemophilia treatment centre to where you will be staying.

Identification cards which contain details of the type and severity of your child's bleeding disorder are available from the National Centre for Hereditary Coagulation Disorders and it is advisable for your child to have this or one of the recognized medical alert symbols with him at all times when he is not accompanied by an adult.

If a babysitter is caring for your child, tell them about his bleeding disorder and make sure that they know who to contact and what to do in an emergency.

Likewise when your child starts playschool make sure his carers are aware of his condition.

WOMEN WITH BLEEDING DISORDERS

Many people think that only men can have bleeding disorders and families are often surprised at the diagnosis of a bleeding disorder in a girl or woman. In girls the first indication that they may have a bleeding disorder is the onset of heavy periods. The most common inherited bleeding disorder which affects both men and women is von Willebrand Disease (see section on von Willebrand Disease)

In very rare cases girls can be born with haemophilia. Some women who are carriers of haemophilia A or B can themselves have lower than normal levels of the clotting factor and can in some cases experience many of the symptoms of haemophilia. For this reason it is very important for known or suspected carriers to know their factor level. Women can also have other factor deficiencies i.e. Factor VII or Factor XI.

Symptoms

Mild bleeding disorders can be much more clinically significant in women. Symptoms of a bleeding disorder can include heavy and prolonged periods, recurring nosebleeds, bleeding from the mouth or gums, easy bruising and bleeding from the digestive or urinary tract. Bleeding often occurs following surgery or dental procedures and following injury.

Women with a severe bleeding disorder are more likely to experience spontaneous bleeding often into joints as happens to men with severe haemophilia.

Diagnosis

Bleeding Disorders in women can remain undiagnosed. It is estimated that:
5% of women age 30 consult their doctors about menorrhagia. (heavy periods)
8% - 10% seeing a gynaecologist have menorrhagia.
10% - 20% of women with menorrhagia if screened, are found to have an underlying bleeding disorder.

When a woman or girl is suspected of having a bleeding disorder it is very important that she be referred to a haematologist who has an expertise in the diagnosis and treatment of bleeding disorders. Due to fluctuation of levels at different stages of a woman's menstrual cycle it is very important to note what stage of the menstrual cycle the blood tests are taken.

CARRIERS

CARRIER DETECTION

The inheritance pattern of haemophilia is a difficult one to understand at first. It is very important to understand it properly so that families who know there is a history of haemophilia can get accurate advice about their own situation.

It is important for female relatives of a person with haemophilia to know their carrier status. They should know about any bleeding tendency prior to surgery or any dental procedure. It is vitally important that they know their carrier status when planning a family.

Obligate Carriers

A daughter of a man with haemophilia.
A mother with more than one son with haemophilia.
A mother with one son with haemophilia and a blood relative with haemophilia.

Possible Carriers

When a man has haemophilia the following family members should be tested for their carrier status:
Mother, Sisters, Grandaughters, Cousins, Nieces, Aunts.

It is important for carriers to be aware of their factor levels as treatment may be necessary at times of severe injury or surgery. When a haemophilia carrier or potential carrier is pregnant she should inform her obstetrician of her carrier status.

It should not be assumed that a young girl who has grown up with a father or brother with haemophilia is aware of her possible carrier status. A young girl needs to be given information relevant to her age and using language that she can understand. This information needs to be given on an ongoing basis as she matures to help her understand it at her level.

When the time comes for a young girl to be tested for her carrier status (usually 16) she will need to be given extra support. Remember that at her age personal image is all important and allow for some negative reaction.

Young girls will be aware of other family members' attitude to the bleeding disorder affecting her family and will be influenced by that.

Genetic Counselling

For carriers or possible carriers it is advisable to undergo genetic counselling preferably before becoming pregnant. Genetic counselling will provide information to enable a woman to make an informed decision in relation to family planning.

Genetic Counselling and Carrier testing is available at the National Centre for Hereditary Coagulation Disorders in St. James's Hospital in Dublin. Tel: 01 4162141

PREGNANCY IN WOMEN WITH BLEEDING DISORDERS/CARRIERS

When a woman with a bleeding disorder or a carrier is pregnant it is vital that there is good communication between the obstetrician, the haemophilia specialist and the paediatrician so that the pregnancy can be managed safely for mother and baby.

In preparation for delivery a plan should be agreed for both the mother and the baby. This should include precautions during delivery, blood samples for diagnosis and the availability of appropriate factor concentrate in the event of bleeding, with instructions on dosage. The plan should be copied to the mother, the paediatrician, the obstetrician and the haematologist.

Remember
Inform your obstetrician of your carrier status or bleeding disorder early in your pregnancy.

WHAT THE IRISH HAEMOPHILIA SOCIETY OFFERS

The Irish Haemophilia Society offers services to all members of the family:

Adults with haemophilia
Women with Bleeding Disorders
Carriers
Parents of children with a bleeding disorder
Kidlink Programme
Youth Programme
Young Adults Programme

The 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.

Information/Education

AGM

The Annual General Meeting and Conference in early March is attended by a large number of people with haemophilia and their families and children.

There are presentations on all aspects of the provision of comprehensive care for people with haemophilia and related bleeding disorders.

There are regular updates on treatments available for those with HIV and Hepatitis C.

A comprehensive educational programme with input from a guidance counsellor, medical presentations and recreational activities is offered for children and young adults.

Creche facilities are provided for infants and very young children to enable parents to attend lectures.

Haemophilia Days

Information/support days for specific groupings within the membership i.e. persons with haemophilia and related bleeding disorders, women with bleeding disorders/carriers, partners, parents, youth group and young adults. These meetings include presentations from relevant medical and other experts on a range of topics related to the particular groups. The meetings also provide an opportunity for mutual support within these groups.

Information meetings are organized on specific topics when appropriate.

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

Counselling /Support Programmes

The counselling/support service is one of the most important aspects of the work of the society.

Members are encouraged to drop into the office at any time if they feel the need to talk to a counsellor.
The society can facilitate members who wish to make an appointment for a definite time and date.
Alternatively for those who cannot avail of the drop in or appointment service our counsellor will arrange to call to their home or to the hospital at a time convenient to them.
The society will respond immediately to a family or individual in crisis and remain until the crisis has been resolved. This service is available throughout Ireland.

Holistic Weekend: The objective of this weekend is to provide support to members with HIV/Hepatitis C their partners and parents and bereaved family members. This popular weekend comprises workshops, relaxation, counselling, and alternative therapies.
Support and assistance is offered to members prior to and during their attendance at the Hepatitis C and HIV compensation tribunal.

The society provides support and education for parents in dealing with a new diagnosis of haemophilia.

This may be an especially hard time for those who have no family history of the condition and we can offer assistance in dealing with medical personnel. Because bleeding disorders are rare conditions

parents may feel isolated and alone and it is very helpful to be put in touch with others in a similar position.

For young members: A programme of support and education addresses the particular needs of this age group in relation to the inheritance pattern of haemophilia and other bleeding disorders.

For students: The Society can arrange help on a number of levels i.e.
Assistance with individual subjects
Assistance with organizing a study plan
Career guidance and help with college applications.

A number of scholarships are awarded annually to students in third level education.

Social activities

Fun days out for all the family are organized each year.

Barretstown Gang Camp allocates places on their excellent Summer Camps, Siblings Camp and Family Camp each year. The Society staff work closely with the staff at Barretstown to ensure that each child gets a chance to experience this wonderful holiday.

Financial Support

Hardship grants can be made available to families in need, especially at Christmas. Assistance can be provided for other needs as they arise.
Membership fees can be waived at the discretion of the Board.
Attendance at the AGM and other Society meetings is subsidized and can be waived in some circumstances.

Services outside Dublin

The headquarters of the Society is in Dublin. The Society is very much aware that members in rural areas often feel isolated. In order to bring the Society outside Dublin staff members visit areas around the country throughout the year.

Hospital Liaison

The Society works closely with all the team members in the National Centre including doctors, nurses, physiotherapists and social workers, many of whom regularly attend Society meetings and weekend seminars. Meetings are arranged in Regional Centres in conjunction with regional visits. Each year at the Annual General Meeting and Conference the Society facilitates a nurses meeting where the nurses from the National and Regional Centres can get together to discuss common issues.

Publications

Four newsletters are published each year with reports on what is happening locally and in the wider world of haemophilia.
The Society produces an annual report outlining the activities and achievements for the year.
Factsheets/Education Leaflets on specific topics are produced as required.
Publications received from sources worldwide are included in our extensive library and are available to members free of charge.

Membership

Ordinary Member

Ordinary members pay an annual fee of €30 and have voting rights. Ordinary members must be 18 years or over. For children under 18 parents will vote in their place.

Associate Member

Associate members are included on the mailing list but do not have a vote. This category can include, other haemophilia organizations, hospital personnel, service providers.

Life Member

These are voting members. They pay a one off fee of €650.

Honorary Life Member

Members who have 30 years continuous membership and who have attained the age of 65 Honorary Life Membership may be given as an award at the discretion of the Board.

Membership Forms available from the Society Lo Call 1850 872 872

SOCIETY REPRESENTATION ON STATUTORY BODIES

National Haemophilia Council (NHC)

The NHC was set up following the Tribunal of Inquiry into the infection of people with haemophilia with HIV and Hepatitis C. The NHC has worked on an ad hoc basis since 2002 and in 2004 the Council was signed into statute.

The membership of the Council consists of:
Two representatives of the Irish Haemophilia Society.
Director of the National Centre for Hereditary Coagulation Disorders (NCHCD)
Consultant Paediatric Haematologist
Consultant Haematologist from outside the Dublin area who treats people with haemophilia
Registered nurse with ongoing involvement in the delivery of haemophilia care
One Health Board representative
One Department of Health representative.

The Council advises and makes recommendations to the Minister for Health on all aspects of haemophilia care. The Council will monitor and make recommendations on specialist services such as orthopaedic and dental services. It will implement and put in place quality management systems and auditing of the services nationwide. The Council can appoint sub groups to carry out specific investigations or tasks on its behalf.

Product Selection Monitoring and Advisory Group (PSMAG)

The PSMAG formally selects the products used for the treatment of haemophilia, von Willebrand Disease and related bleeding disorders from all the products available.

The membership of the PSMAG consists of:
Two representatives of the Irish Haemophilia Society
Three Clinicians
One Department of Health representative
One Eastern Regional Health Authority representative
One Irish Medicines Board representative
One virologist
Two representatives of the I.B.T.S.
Two outside advisors (one nominated by the I.H.S.).

The functions of the PSMAG are as follows:
To decide on the products to be purchased on a national tender basis
To ensure that products purchased meet the agreed criteria
To tender, place contracts and administer contracts.
To monitor product delivery
To put in place emergency backup procedures in relation to ongoing safety, supply and availability
To monitor international developments
To monitor the evolving risk of infection and take appropriate action to minimize risks.
The Society through its role on the PSMAG will continue to monitor international developments and respond to any future perceived threat to the safety and/or supply of all products used for the treatment of bleeding disorders in Ireland.

Hepatitis C Consultative Council

The Hepatitis C Consultative Council was established by the Minister for Health and Children in 1996 to advise and make recommendations to the Minister for Health on the organisation, delivery and confidentiality of services for people who contracted Hepatitis C from contaminated blood products. Members of the Council are appointed by the Minister for Health for a three year period. The Irish Haemophilia Society is represented on the Council. The Council can appoint sub groups to carry out specific tasks in relation to any aspect of the services for people with Hepatitis C.

Further information on the Hepatitis C Consultative Council is available from the Society Lo Call 1850 872 872 or on the Council's website www.cchepc.ie