HAEMOPHILIA & VON WILLEBRAND DISORDER: AN ESSENTIAL HANDBOOK FOR PARENTS

IRISH HAEMOPHILIA SOCIETY
REPRESENTING PEOPLE LIVING WITH HAEMOPHILIA, VON WILLEBRAND'S AND OTHER INHERITED BLEEDING DISORDERS

Contents

Introduction

As a parent, when a child is diagnosed with a bleeding disorder, you may feel scared, worried and perhaps even guilty. 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 and concerns. After a while, you will find that it is not nearly as bad as they first imagined. The impact of replacement therapy factor concentrates or novel threapies on the lives of people with haemophilia needs to be explained. It is important to remember that with good treatment the child with a bleeding disorder has every chance of growing up as an active, fit person 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 Irish Haemophilia Society can bring families together through education, publications, conferences, information meetings, regional visits, home and hospital visits and outreach. The Society offers support and advice to help improve the quality of life for people with bleeding disorders.

Bleeding Disorders

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. Blood contains many proteins called clotting factors, which work to stop bleeding. The lack of clotting factor causes people with haemophilia to bleed for longer periods of time than people whose blood factor levels are normal. However, people with haemophilia do not bleed faster than other people.

Most bleeding in haemophilia occurs internally, into the joints or muscles. The joints that are most often affected are the knee, ankle and elbow. Repeated bleeding without prompt treatment can damage a joint.

They don't have enough clotting factor VIII

Affects 1 in every 5,000 males

They don't have enough clotting factor IX

Affects 1 in every 30,000 males

There are two types of haemophilia. Haemophilia A is a deficiency in factor VIII and haemophilia B (sometimes called Christmas Disease) is a deficiency in factor IX. Both types of haemophilia share the same symptoms and inheritance pattern and 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.

Severity of haemophilia	Percentage of normal factor activity in blood	Number of international units (IU) per millilitre (ml) of whole blood
Normal range	50%-150%	0.50–1.5 IU
Mild haemophilia	5%-40%	0.05–0.40 IU
Moderate haemophilia	1%-5%	0.01¬–0.05 IU
Severe haemophilia	less than 1%	less than 0.01 IU

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

Haemophilia is a genetic blood disorder, which is usually inherited although in 30% of cases, there is no family history and the haemophilia is caused by a spontaneous mutation in the X chromosome. The gene is passed down from parent to child. A person who is born with haemophilia will have it for life. Some of the signs of haemophilia are: large bruises, bleeding into muscles and joints, spontaneous bleeding and bleeding for a long time after a cut or surgery.

von Willebrand Disorder

von Willebrand Disorder (vWD) is the most common type of bleeding disorder. People with vWD have a problem with a protein in their blood called von Willebrand Factor (vWF) that helps control bleeding. When a blood vessel is injured and bleeding occurs, vWF helps cells in the blood, called platelets, mesh together and form a clot to stop the bleeding. People with vWD do not have enough vWF or it does not work the way it should. It takes longer for blood to clot and for bleeding to stop.

vWD is generally less severe than other bleeding disorders. Many people with vWD may not know that they have the disorder because their bleeding symptoms are very mild. For most people with vWD, the disorder causes little or no disruption to their lives except when there is a serious injury or need for surgery. However, with all forms of vWD, there can be bleeding problems.

It is estimated that up to 1% of the world's population has vWD. Many with mild vWD will not require treatment. Research has shown that as many as 9 out of 10 people with vWD have not been diagnosed.

There are three main types of vWD. Within each type, the disorder can be mild, moderate or severe. Bleeding symptoms can be quite variable within each type depending, in part, on the vWF activity. It is important to know which type of vWD a person has because treatment is different for each type.

Type 1 vWD is the most common form. People with Type 1 vWD have lower than normal levels of vWF. Symptoms are usually very mild. However, it is possible for someone with Type 1 vWD to have serious bleeding.

Type 2 vWD involves a defect in the vWF structure. The vWF protein does not work properly, causing lower than normal vWF activity. There are various Type 2 vWD defects. Symptoms are usually moderate.

Type 3 vWD is usually the most serious form. People with Type 3 vWD have very little or no vWF. Symptoms are more severe. People with Type 3 vWD can have bleeding into muscles and joints, sometimes without injury.

Von Willebrand's Disorder affects both men and women. However, because vWD can cause heavy menstrual bleeding and prolonged bleeding after childbirth, more women than men have noticeable symptoms.

Rare Bleeding Disorders

Rare clotting factor deficiencies are bleeding disorders in which one or more of the other clotting factors (i.e. factors I, II, V, VII, X, XI, or XIII) is missing or not working properly. Less is known about these disorders because they are diagnosed so rarely. In fact, many have only been discovered in the last 40 years.

Diagnosis

Family History

If a family has a history of haemophilia, baby boys will usually be tested soon after birth. Even when this does not happen, babies with severe haemophilia are often diagnosed early in life, because they show signs of bruising or bleeding when they begin to crawl or walk. Boys who have moderate or mild haemophilia may not show such obvious signs of haemophilia. Often the first sign in these boys is bleeding after removal of teeth or following surgery or a serious injury. Some people with mild haemophilia may reach adulthood before they are diagnosed.

New Diagnosis

Many boys with haemophilia are diagnosed soon after birth, but those with a mild form of the condition may not be diagnosed until they are much older. This section describes how a diagnosis is made and what services are available to affected boys and their parents and families. Common, natural reactions to what may be an initially shocking or upsetting diagnosis and ways of coping with these reactions are also described in this section.

How haemophilia is diagnosed

Haemophilia can be diagnosed by testing a blood sample for its factor VIII or factor IX activity respectively for haemophilia A or B. This activity can then be compared with activity in people known not to have haemophilia. Haemophilia is divided into three categories according to a person's factor VIII or factor IX activity:

- Less than 1% of normal factor VIII / factor IX activity results in severe haemophilia
- 2-5% of normal factor VIII / factor IX activity results in moderate haemophilia
- 5-40% of normal factor VIII / factor IX activity results in mild haemophilia.

Coping with the diagnosis

Receiving a diagnosis of haemophilia is naturally a stressful event, after which parents anxiety increases. It is common for parents to experience a grief like reaction. This grief reaction involves a range of emotions in no particular order, including, shock, sadness, denial or disbelief, anger, guilt and eventually acceptance. If your family has no previous history of haemo-

philia, the diagnosis will obviously come as a shock. Even if there is a family history, receiving a diagnosis can be difficult.

There is a lot of new information and a whole new medical language to take on board. The period after initial diagnosis can be quite challenging. Adjusting to and coming to terms with your child's condition takes time and is a process not an event. Getting accurate information about your child's condition will help give you a sense of control. It is important to take time to think about how best to cope with strong feelings. Staying in contact and getting support from close family and friends is important. Everybody copes differently, we can learn from what helped us deal with stressful situations in the past. Many people benefit from accessing emotional support from professionals such as psychologists, counsellors or social workers. Acknowledging how we are feeling and expressing our emotions can be helpful as parents adjust to the new demands of their child's condition. A diagnosis can be particularly hard for anyone who has not come across this condition before. If this is the case, the diagnosis may be frightening, and you might feel that your son needs to be protected and treated differently from other children. However, you may find it helpful to bear the following in mind.

- Your son is a baby first, and a baby with haemophilia second. He is the same baby he was before the diagnosis was made and needs just as much loving as before.
- You will not cause your son any harm by doing the usual things, such as kissing and cuddling him.
- Your son will grow up normally and be able to live a normal life.

Although haemophilia is a serious condition, treatments and the long-term outlook for people with haemophilia are improving all the time.

Once you have got over the initial shock of the diagnosis, you should start to think about your hopes and fears for your son and discuss these with the experts at the haemophilia centre. Common questions usually cover subjects such as:

- How haemophilia will affect your son's health?
- What symptoms he will have?
- How your son has inherited haemophilia, and what this means for the rest of the family?
- What treatments are available?
- When you should consult your GP, and when to go to the haemophilia centre?
- How haemophilia will affect your son's schooling and employment prospects?
- Which sports are safe for him to participate in?
- Whether it is safe for him to travel

Obviously, you will not want answers to all these questions at once, and you can talk to the staff at the haemophilia centre anytime during your son's upbringing to discuss issues as they arise.

What services are available following diagnosis

Now your son has been diagnosed as having haemophilia, the following services should be provided by the haemophilia treatment centre.

- He should be given a full medical examination to ensure that he is in good health.
- The doctors should tell you the most appropriate way to manage your son's haemophilia, including any treatments that will be prescribed and regular check-ups to be arranged with the haemophilia centre.
- You should be told about the role of the haemophilia centre, both treatment and in counselling him and your family.
- You should be given information on all the members of the comprehensive care team at the haemophilia centre, and how to contact them during office hours.
- You should also be given full details of who to contact and how to get ambulance transport in an emergency, and how to get help from the haemophilia centre outside of office hours. This information should be passed on to anyone likely to be looking after your son.

Treament

Types of Treatment

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. Some children self infuse. Bleeding can be internal or external.

Treatment for bleeding into the joints and muscles:

- Coagulation factor concentrate (IV),
- R.I.C.E. - (Rest, Ice, Compression & Elevation)

Prophylaxis

It is recommended that all children with severe haemophilia who are over two years of age or who have had two joint bleeds receive regular doses of coagulation factor concentrate to prevent spontaneous bleeding and reduce the risk of long-term side effects. This is called prophylaxis. For children with severe factor VIII and factor IX deficiency, this involves an injection of coagulation factor concentrate into the veins on a regular basis, usually one to two times per week. In Ireland, the factor concentrates currently used are extended half life factor concentrates which are infused less regularly then regular factor concentrates and offer more protection against bleeding episodes. In small children and children with poor veins repeated intravenous injections can be very difficult and a port-a-cath or 'port' may be needed until the veins have developed. By the time your child is ready to start prophylaxis, you will have discussed intravenous access and ports with your haemophilia team.

For more information on Ports/PICC lines etc. please see Appendix 1 on page 34.

Subcutaneous Therapy

From late 2019, a new generation of treatment for people with severe factor VIII deficiency is available as an option in Ireland. This treatment, called Hemlibra, is a monoclonal antibody which mimics the effect of factor VIII. It can be infused weekly or every 2 weeks subcutaneously (under the skin) thereby avoiding the requirement for regular intravenous injections. This treatment offers a high degree of protection against bleeding but can not be used to treat bleeding episodes. In the event of a bleeding episode, intravenous infusion with FVIII concentrate will continue to be required. This subcutaneous therapy can not be used for factor IX deficiency so , in this case , the routine treatment will continue to be extended half life factor IX concentrate.

Self Infusion

Your child can learn to self infuse. Yes, it can be something you do yourself. Training can be arranged by contacting the haemophilia team at Children's Health Ireland at Crumlin.

Items needed for self-infusion:

- A tourniquet
- Alcohol wipes
- Butterfly needle
- Tape
- Syringe
- Factor concentrate
- Sharps container

Preparation:

- Make sure your child's hands have been washed and the area is clean.
- The relevant limb should be adequately supported on a stable surface below heart level to allow gravity to enhance venous dilation. This causes blood to pool and veins to distend. Lightly tapping or gently stroking the vein along its length in a downward direction will also cause venous distension. Another simple, effective way to dilate veins consists of alternately clenching and relaxing your fist.
- Feel the veins to see how big they are, and in which direction they go, choosing ones that are straight. Start by placing one or two fingers over the vein and press lightly. Your thumbs should not be used as they are less sensitive.
- Apply a tourniquet two inches above the potential site for one minute only. Allow a few minutes for the vein to become more prominent.
- Select the vein, clean the skin with an alcohol swab in a downward stroke, and allow skin to dry.
- With the tourniquet applied, the vein identified, and the factor ready, hold the butterfly wings and insert the needle into the vein with the bevel facing upwards.
- When there is blood flashback (visible in the butterfly tubing), remove the tourniquet and inject the factor concentrate slowly.
- Once this is complete you can withdraw the needle, making sure to put pressure over the vein with a piece of cotton wool for two or three minutes. Then place a plaster on the site.

For more information please contact the haemophilia team at Children's Health Ireland at Crumlin.

Inhibitors

People with haemophilia receive factor concentrate to help their blood to clot. However, in some people, the immune system will then start to produce antibodies to block the effects of the treatment. These antibodies are known as 'inhibitors'. The inhibitor can make the medication used to treat haemophilia less effective. This means bleeds become more difficult to control. This can lead to more bleeding into muscles and joints resulting in more joint damage and pain. When this happens, more intensive or alternative treatment may be required to control bleeds or to attempt to eliminate inhibitors. Most inhibitors are temporary. They most often appear during the first year of treatment but they can appear at any time.

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.

Inhibitors of factor VIII occur in about 20% of individuals with factor VIII deficiency. The inhibitors can be high titre or low titre. High titre inhibitors refer to a higher level of inhibitors which is more clinically significant and more difficult to eradicate. Low titre inhibitors are often transient and the person can still be treated with factor concentrates at higher doses. If a child develops a high titre factor VIII inhibitor, the optimum therapy is immune tolerance therapy where the child is treated with very high doses of factor VIII for an average period of about one year, in an attempt to eradicate the inhibitors. Treatment for factor VIII inhibitors can be with a number of agents including a recombinant factor VIIa (Novo Seven) or an alternative product known as FEIBA.

From 2018, a new generation of treatment for people with factor VIII inhibitors has been used in Ireland. This treatment, called Hemlibra, is a monoclonal antibody which mimics the effect of factor VIII. It can be infused weekly or every two weeks subcutaneously (under the skin) thereby avoiding the requirement for regular intravenous injections. This treatment offers a high degree of protection against bleeding but can not be used to treat bleeding episodes. In the event of a bleeding episode travenous infusion with bypassing agents (usually recombinant factor VIIa) will continue to be required.

Inhibitors in patients with factor IX deficiency are extremely rare and are more difficult to treat. In general, children and adults with haemophilia are screened for inhibitors at least once or twice per annum.

Bleeds

Bleeds in infancy

Identifying a bleeding episode during infancy can be very challenging for parents. During the first six months of life, bleeding episodes are minimal. Superficial bruising can be commonly seen on the chest or back from picking up the infant. These bruises are usually minor and do not require treatment. They can be minimized by cradling the infant when picking him up.

General guidelines for administering immunizations include using the smallest gauge needle possible. An ice pack can be applied for five to ten minutes after the injection to help decrease the risk of bleeding. Also, place a barrier such as a gauze or a washcloth between the ice pack and the injection site so the ice will not be in direct contact with the skin. Immunizations commonly cause redness, warmth and minor swelling at the site of injection. Infants can also become irritable following immunizations. Children with haemophilia should be observed carefully for signs that could indicate a bleed.

If you observe:

- Your child is very irritable
- He guards or will not move the area where the immunization was given
- Redness, warmth or swelling increase

You should:

- Apply an ice pack to the site
- Call your clinic if symptoms persist

During these first few months it is important to observe your infant for signs of bleeding that may require treatment. "Body checks" during diaper changes and bath times are an excellent opportunity. Lay your baby down and observe movement of his arms and legs.

Does he move them freely? Talk to him and hold a toy for him to reach out to. Will he do it? If he is not moving an arm or leg, investigate why. If a toy is moved close to his arm, will he reach out? If not, move the arm gently yourself. Does he grimace, cry, or hold it stiffly? Does one arm or leg appear larger than the other? If so, it could be the sign of a bleed. Is there any swelling or bruising, or is the area warm to the touch?

Contact your clinic staff to discuss your observations. Be prepared to describe:

1. Where you suspect the bleed is
2. What the area looks like; is there bruising?
3. Location and size of swelling
4. Does it feel warmer than the surrounding area?
5. When you first noticed the bleed?
6. Is it getting worse?

Bleeds in preschool years and upwards

Children at this age are more coordinated. Their height, weight and physical activity are increasing as they explore more of their environment. It is at this age that more muscle bleeds can occur, and joint bleeds begin. Some system of body checks should continue throughout this period. These body checks can occur when he dresses in the morning or at bed or bath times. Involve your child in this process so he can begin to learn what to look for. Encourage him to tell you when he is having a bleed.

Signs of a muscle bleed

- Tenderness to touch
- Swelling
- Warmth
- Pain with movement
- Muscle feels "tight"

Signs of a joint bleed

- Early tingling
- Feels "funny"

Followed by:

- Pain
- Warmth
- Swelling
- Stiffness
- Limited movement

Muscle and joint bleeds should be treated with factor as early as possible to prevent muscle and joint damage, followed by rest, ice, compression and elevation of the joint:

Rest:

Ice:

Compress:

Elevate:

Remember to:

- Praise your child for his responsible behaviour when he tells you of a bleed.
- Emphasise the importance of early treatment; maybe only one treatment instead of several.
- Reinforce that infusions are not punishments but rather treatment to make what hurts better.

Joint & muscle bleeds

For those severely affected, a major problem can be 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. 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 normal everyday activity. These bleeds are sometimes described as "spontaneous" because it is impossible to identify a cause.

An ache or irritation in an affected area is usually an indication that a person with haemophilia is getting a bleed. If left untreated pain may become 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 ankles and knees being the worst affected in most people.

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 ones may seem upset or may protect a limb by limping or not using it.

Cuts and grazes

When cuts and grazes occur, 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.

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 treatment centre but try giving the child an ice cube or ice pop to suck as this may do the trick.

Soft Tissue Bruises

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 size and is causing pain, then treatment may be recommended.

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 can result from a severe bang 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 should be taken to the nearest haemophilia centre.

Recognising a bleed

Signs of a head bleed

- Nausea and /or vomiting
- Lethargy & unsteady balance
- Irritability & confusion
- Headache & drowsiness
- Seizures
- Loss of consciousness

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

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

A CAREFUL EYE SHOULD BE KEPT ON THE CHILD FOR AT LEAST 12 HOURS FOR ANY OF THE SYMPTOMS LISTED.

Signs of a mouth/nose bleed

Any bleeding lasting longer than 20mins from mouth, gums or throat

Signs of an Abdominal Bleed

- Bloody, black or tar-like stools
- Red or brown urine

Signs of a joint or muscle bleed

- Pain, Warmth, Swelling,
- Tingling/Tightness, Tenderness
- Reluctant to move affected limb

Pain

Types of pain

Pain associated with haemophilia and other bleeding disorders is usually of two types:

- Acute pain is short-term. It's caused by bleeding into a joint or muscle and is usually the earliest sign of a bleed. A bleed that is not treated quickly can make the pain much worse. Even if there are no other obvious signs of bleeding, such as swelling, factor concentrate is recommended.
- Chronic pain is longer-term. It's caused by long-term, or repeated, joint damage and typically affects older people with haemophilia who have 'built' up joint damage over the years.

Dealing with pain at home

It is important to treat bleeding episodes without delay as this will minimise the pain caused by internal bleeding into joints or muscles. Untreated bleeds result in excruciating pain and arthritic deterioration. Long term arthritic deterioration can lead to constant pain in a damaged joint.

Pain from a bleed can be managed by giving factor replacement infusions to stop the bleeding. There are also non-medical techniques that can be used. Applying ice may also reduce acute pain. Ice should not be applied directly on the skin. Use a towel and do not apply ice for more than 10 minutes at a time. Supporting the arm or leg on a pillow, or in a splint, may also help. A relaxing activity can help soothe muscles, and take someone's mind off their pain.

Chronic pain can be very difficult – and significantly affect a person's quality of life. But there are a number of treatment options available. Be as specific as possible with your HTC team about the pain you're feeling, so they can identify possible causes and suggest solutions. A consultation with a physiotherapist can also be helpful. He or she may suggest different strategies to help manage the pain, including exercises and mobility devices. Talking to your teams paediatric psychologist may also help as they can support and advise you and your child on strategies to help you and your child cope with the emotional aspects of pain – which are often forgotten, but important to deal with.

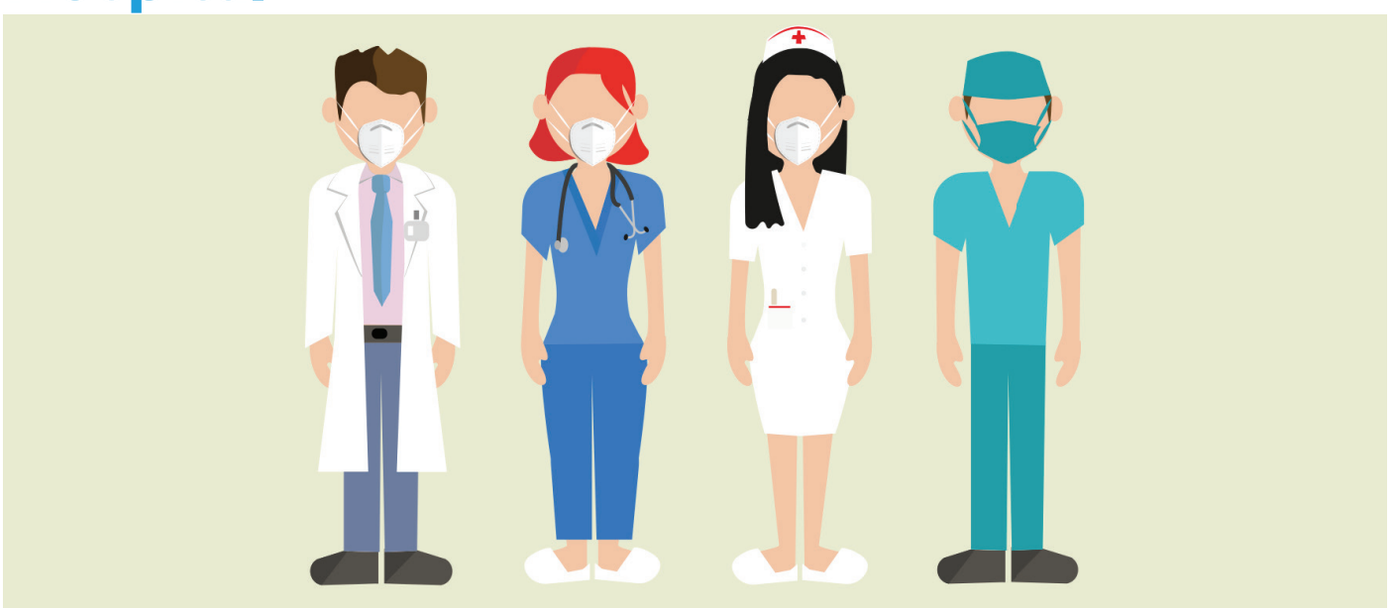
Medications to avoid

- People with bleeding disorders should avoid aspirin and other drugs that can cause bleeding. Many non-prescription medications, such as cold remedies, contain aspirin. People with bleeding disorders should check with their haemophilia treatment centre before taking any new medication.
- People with bleeding disorders should check with their haemophilia treatment centre before taking any new herbal drug.

When to go to the hospital

If your child is receiving regular factor infusion treatment and it does not seem to relieve the pain, or if a bleed seems to be getting worse, go to your nearest haemophilia treatment centre straight away. It's possible that inhibitors have developed that are interfering with treatment, or there may be another reason for the pain that's not related to bleeding.

Hospital



Getting ready for the hospital

Helpful hints getting ready for the hospital

- Keep a 'clinic bag' ready for those trips to the hospital. Include a special game for your child to play with.
- Keep instant ice packs on hand to use in a hurry, when travelling, or for your child's school to keep.
- Gel ice packs work especially well – they are reusable and stay flexible.
- Keep an extra soother in the freezer. It's great for bleeds when cutting teeth.
- Keeping children off their feet while bleeds resolve can be a challenge.
    - Read to them. Get them interested in books.
    - Keep several puzzles on hand.
    - Play games with them.
    - Stay in their view so they won't be tempted to look for you.
    - Rest with them. You probably need and deserve it.

Involve your child as much as possible in decisions that involve his treatment. The older the child, the more decisions he can make:

- Will he sit or lie down?
- Where will the stick with the needle be?
- Have him help get infusion supplies ready
- Will he hold a special toy or squeeze your hand?

Clinic Appointments

- It's always a good idea to arrive early for a clinic appointment.
- If this is your first clinic appointment a full medical history of your child will be taken by the consultant.
- Bring a record of any medications your child is currently taking.
- Please also remember to bring vaccination records for your child.
- And don't forget its always a good idea to write down any questions you might have before the appointment so that you are prepared.
- If you are unsure of some of the medical jargon that is being discussed, don't be afraid to ask questions.

Accident and Emergency Department

Bleeds can often occur in the evening, or on weekends or holidays. It can help to ensure that your child gets the proper treatment in a more timely fashion. Emergency Room staff may not be familiar with your child or haemophilia. This is an important reason why parents need to be advocates when it comes to their child's care.

Be prepared to:

- Clearly express your child's needs
- Accurately describe the bleed that needs to be treated
- Know what type of haemophilia your child has
- Be able to name the product he is treated with
- Check to be sure he receives the correct dose

A&E rooms treat patients on a priority basis. They may not be aware that prompt treatment of bleeding episodes is important in preventing complications that can occur from prolonged bleeding in persons with haemophilia. It is not unreasonable for an inquiry to be made if your child is not seen within 30 minutes. Remind the staff of the importance of prompt treatment and ask how much longer it will be. Be understanding of the A&E situation as you calmly and clearly express your needs.

Ensure you have a "Severe Bleeding Disorder Alert" card for your child which you always carry with you. This is available from the Haemophilia centre at Childrens Hospital, Ireland in Crumlin. If you need further information about this card, please contact the society.

Emergencies in your absence

Accidents can happen at any time. It is important to plan ahead so others will be able to handle emergencies in your absence.

- Inform your child's caregiver of his bleeding disorder and what to do in the event of a minor injury.
- Be specific as to how major injuries should be handled.
- Post emergency information near the phone. Instruct the caretaker to take this information if they go for treatment.
- Emergency information should include:

    - Where you can be reached
    - Name of your child's doctor
    - Type of haemophilia and usual treatment
    - Clinic or emergency room number

- If factor is stored at home, be sure instructions are clear as to where it is kept and the amount that should be taken.

Hospital admissions

Your child's hospital admission can affect many aspects of your family life, be it following a trip to the Accident & Emergency Department or in relation to a planned admission. You may experience a wide range of emotions. These emotions can range from worry and fear to relief. By recognising these emotions you will be in a better position to support your child. So it is just as important to prepare yourself as it is to prepare your child. Children often sense their parents feelings. By recognising your own fears, worries etc it will allow you to be in a better position to support your child during his/her admission to hospital and cope with any stresses that may arise.

Hospital admission can be stressful for parents and children of all ages. Preparing yourself and your child in advance may help reduce some of the anxiety and allow you and your child to cope well. Generally, providing age-appropriate, clear, accurate and honest information is advised. Children tend to cope best when they know what to expect. How long in advance you prepare them will vary depending on the age and disposition of your child. Younger and more anxious children are best informed of hospital admissions closer to the time so as to avoid too long of a lead and too much time to worry.

Younger children generally need to have a parent stay with them to explain, support and comfort them during their hospital stay. Preparing your child is more than giving them information. It is understanding and responding to your child's individual reactions. It is important to get all the information you can about your child's hospital stay. Having a clear picture yourself will empower you to answer most of your child's questions. Ask the doctor or nurse what tests and procedures your child will need while in hospital, how long is the expected admission?

There is no right or wrong way for children or families to deal with a hospital admission. However, in general the more a child knows in advance the more comfortable he or she will be. No matter what age your child is the following tip may help:

Talking to your child:

Each child is unique and you know your child best. Choose a quiet time to talk about the upcoming admission or surgery. Give information in age-appropriate language, using your child's words. For younger children you might find it helpful to use a story book format explaining what will happen and when, using a beginning, middle and end. Emphasise the positive parts about having the procedure and acknowledge the annoying and painful parts. Speak slowly, using a calm, relaxed voice. Talk about the hospital admission in a cheerful, matter of fact, positive way explaining that doctors and nurses are here to look after him/her. Very often it is not what you say but how you say it, if you are calm and positive your child is more likely to be calm and positive. Repeat and summarise what you have explained and check that your child has understood it. Reassure your child that the admission, operation, test or procedure is necessary and the right thing to do. Planned (non-emergency) admissions can often be very confusing to a child as they feel well and may not understand why this admission is necessary. Give your child clear, honest concise information.

Growing up with a Bleeding Disorder

Infants & Toddlers

Because most bleeding disorders are genetic, there is often a history of the disorder present in the family. This is despite the fact that current treatments are very safe and effective and quality of life for people with haemophilia has improved dramatically. On the other hand, approximately 1/3 of all bleeding disorders diagnoses are a result of spontaneous mutation.

Infant and toddler years can be both the most wonderful and most stressful time in your child's developmen. Parents must learn to deal with first bleeds and treatment that seems invasive and traumatic for their child. Toddlers live in the moment and their natural curiosity and drive to explore mean this stage is particularly difficult for parents. Toddlers require constant vigilance. It is difficult to strike the balance between allowing your toddler explore and experience the world and keep them safe at the same time. Teething also begins at this time and causes children to chew on objects to seek relief, thus resulting in a greater potential for mouth bleeds. Although this is a normal and necessary part of his development, mouth bleeds mean additional emotional distress for you. This type of bleeding can be very difficult to stop due to the fact that the mouth is warm, wet and in use so often. Hospitalisation may even be necessary. Later, when the child begins to pull up and walk, more bleeds begin to occur.

Parents' natural response at this time is to increase their protectiveness in an attempt to prevent bleeding episodes. Those who have a family history may slide into memories of their past experiences with the bleeding disorder and react by restricting their child just like in the "old days". Some parents resort to carrying the child most of the time or keeping him confined in a high chair or playpen. Yet these restrictions then tend to stifle children's natural instincts and development / if children are denied the opportunity to explore and manipulate objects, they are likely to become timid, fearful, and have trouble becoming self-reliant. The conflict between wanting to meet their child's social and emotional needs and physical safety needs places parents under a great deal of strain regardless of the severity of the child's bleeding disorder.

Parents of children with bleeding disorders often find it difficult to make time for themselves and/or each other.

There are several reasons this may be true:

1. It may be difficult finding capable and responsible babysitters who comfortably agree to care for your child, knowing they must invest time to learn about your child's bleeding disorder.
2. Extended family members are often uncomfortable caring for the child with a bleeding disorder, especially when there is no family history of the disorder. Many parents report that aunts, uncles, cousins and grandparents will only say "yes" to caring for the child briefly if at all, citing fear and anxiety in assuming responsibility for the child as the primary reason for saying "no".
3. Many parents report feeling a great deal of anxiety when leaving their child with anyone else. They most fear a bleed occurring or perhaps being overlooked or mishandled in their absence.

Despite these difficulties, it is important for children to have the experience of their parents being absent as well as present on occasion during the first three years of their lives. Being left in the care of other responsible adults and with other children builds an important foundation in your child's socialization experience, trust-building process and security base. Developing the security of knowing their parents can leave them and return is important for forming this base.

Although it may be difficult to arrange, and you may feel uneasy about leaving your child at day care or in the care of others, it is important in his early development to make every effort to do so, even if only one or two times a month. Choosing one or two regular people or places to care for your child is advised in order to promote continuity and regularity for him. Remember that trust develops from your child's perception that his surroundings and caretakers are confident, consistent, nurturing and supportive. Therefore, minimising your child's perception of the negative effects of his disorder by treating it as a "part of life" will help him learn to trust in your protection and ability to take care of him. Later he will begin to learn to trust in his own ability to take care of himself.

While it is natural during this period to want to restrict you child to a playpen, children with bleeding disorders, just like other children, need to be allowed to explore. They need to experience all that other children their age experience, such as crawling on the floor, holding and touching objects and playing with siblings. They also need to know what it is like to be left with a responsible babysitter, and to be held and loved by parents, grandparents and older siblings. They need to learn to accept "no" to some of their demands, and they need to interact with family and neighbourhood children. Keeping in mind that you want to give your child the highest level of understanding and comfort in living "normally" with a bleeding disorder, you can help him develop trust and self-esteem by finding a balance between safety needs and emotional needs. Allow him to explore as much as possible while taking reasonable safety precautions at the same time.

Early childhood, ages 3 to 6

These early years are a wonderful and exciting time, seeing your child's personality emerge. With language development you get a window into their thoughts. It can also be very anxious time as you try to balance their continued need to explore with their safety and well-being.

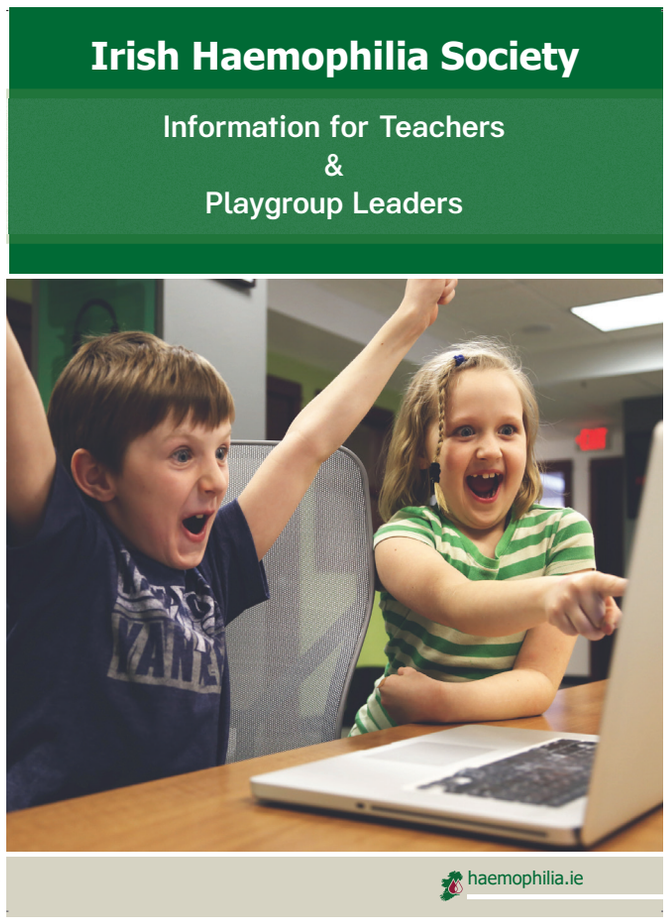
The increased frequency and severity of bleeds resulting from toddlers becoming mobile heightens disruption to family life. There is also a greater requirement for accurately assessing the need for treatment. Parents must learn more about their child's particular bleeding disorder. Children become more able to express their discomfort from bleeds and treatment. Prophylaxis, or regular treatment to prevent bleeding episodes will usually be commenced by the age of 2 which gives more protection to the active toddler.

These factors usually result in parents introducing more restrictions on activity. Again, finding a balance between safety needs and social and emotional needs is essential. At the same time your child will usually be able to begin to take part in his treatment by "assisting" the nurse. He can open the alcohol swab packets and choose his Band-Aid. He can also take more control over infusions by cooperating. If you continue to treat these issues objectively, your child is much more likely to cooperate and to have more positive experiences. You will be setting the stage now for how well your child is able to deal with emotional aspects of his bleeding disorder in the future. Acceptance of the bleeding disorder as a fact of life and developing a matter-of-fact attitude is important because there is much more to come. Children are resilient. Expect appropriate behaviour from your child, generally speaking, parenting and discipline should be the same for children with haemophilia as for those without. Building a positive relationship with your child and helping them to develop a good understanding and acceptance of their haemophilia will be hugely beneficial as your child grows.

Parents who have put their careers on hold may now want to return to work, so the issue of finding a day care centre that will accept their child may arise at this time. As discussed earlier in his class, owners of day care centres often fear problems with liability if the child has an injury on their premises. Day care centre staff has little understanding of bleeding disorders and because the disorder is relatively rare, child care staff may have little motivation to learn more. An information booklet

is available from the Society for Teachers and PlayGroup leaders

The treatment centre staff will often be willing to visit the facility to help with your child's placement. At this time, parents wishing to return to work will have their first experience of advocating for their child with the help of the treatment centre staff. If you have not already faced this problem in the past, you may now feel uneasy about allowing your child to go to preschool or day care. Parents are often fearful of a bleed being overlooked or mishandled. It is important to deal with your own feelings of protectiveness when it is time for your youngster to be left in the care of others.



School age, age 6 to 12

In terms of treatment, this age is usually the busiest. A lot will happen in the next three years. For instance, home-infusion training, or immune-tolerance therapy may be started. This could involve the insertion of a port-a-cath. The availability of subcutaneous therapy for factor VIII deficiency will remove the possible need for port-a cath insertion.

With the growing language development and body awareness that occurs during this period, children can now tell their parents when they have a bleed. Be aware that they may also choose not to tell because they know the bleed may require a trip to the treatment centre or home treatment that may interrupt an enjoyable activity. It is common during this stage of development for children to start asking questions about haemophilia. Questions such as "why do I have haemophilia?", "will I always have it?" are common.

Some boys may complain about it and say they "hate it". This is all very normal and part of the process of your child's growing understanding of their medical condition. It is important to let them vent these feelings, acknowledge them and help your child in acceptance of his haemophilia.

Keep in mind that people tend to fear the things they cannot control. It is important to show children that they have control over their disorder. This can be accomplished by including your child in the infusion process.. Your child may now be able to help the nurses arrange supplies for the infusion and helping to find veins. They can also help by putting on a tourniquet and selecting Band-Aids.

Adding to the stress of this period for parents is the increased necessity to assess bleeds accurately. Since your child is now able to express pain much more accurately, as a parent you must seal with your own emotions in regard to your child's discomfort. Your child may want to avoid treatment because he does not want an infusion. In many respects, parents are caught in a conflicting situation. If they go to the HTC when it is unnecessary, they create a heightened sensitivity to the child's disorder for the whole family and increase the child's pain. If they fail to obtain treatment when it is needed, the bleed may become worse and require additional days or weeks of infusions. A balance between overreaction and accurate assessment is often a trial and error process. As you increase your knowledge about your child's disorder, it will become easier to maintain normality in your home. The importance of a little boy's relationship with his father at this age cannot be emphasised enough. However, fathers with children at this age often begin to realise how the bleeding disorder may impact their dreams of having a son with whom they can play various contact sports. If it has not begun to occur already, it is at this point for these men that their disappointment regarding limitations on such play may begin to affect their desire to build a relationship with their son. Learning ways to play with their child in new and equally relatable ways will set the stage to forge their new and satisfying relationship from refocused dreams. The benefits to the child of his father doing this will last a lifetime.

Late in this period, children will also begin to lose baby teeth. In some families, the joyous anticipated arrival of the tooth fairy may be clouded by mouth bleeds that can occur from this normal occurrence. Patience, understanding and contact with the treatment centre make this stage easier to deal with.

Teenagers

The teenage years can be a difficult developmental period where young people struggle to become individuals. It is a difficult time for parents too! You probably remember being a teenager – and testing the boundaries with your parents. Your child may do the same as they enter their teen years. Some may take more risks than others. Striking the balance between laying down the rules and not damaging the relationship with your teenager can be tricky. It is important to try and keep communication open. Pick your battles carefully, there's only so much you can do as a parent – if your kids test the limits, and push back, it's something you'll have to deal with together. What's key is that taking responsibility for their health remains at the forefront of your child's mind.

Teenagers with haemophilia usually start to make more of their own decisions regarding the activities they choose and their treatment. Peer pressure can be especially difficult for a teenager who feels 'different' because they have haemophilia. This may lead to more risk-prone decisions, such as participating in higher-risk sports, like gaelic football, hurling or rugby. As a parent, it's important to have a frank discussion with your teen about the potential risks of the activity, and the protective measures (such as special equipment) needed. Together, you can make a decision that helps your teen grow and bond with their friends and minimize risks. As a reference, you can use the 'Sport and Exercise' publication from the Irish Haemophilia Society.

As teens start to take more responsibility in managing their own care, one of the biggest areas of growth may be learning to self-infuse. This involves the technique itself, plus organizing appointments and follow-up and sticking to the treatment schedule.

As a parent, allowing your teenagers to start making their own decisions, and depend less on you, can be a difficult transition. It can be hard letting go. Again, having an open dialogue with your teen about what it means to manage your own care, and the consequences of not staying vigilant, is important. Also remind them that you'll always be there for support, whenever they need it. As your teen starts taking on more responsibility, he'll become more confident – which, in turn, will give you confidence that he'll be ok.
Teenage years are a time of great physical, emotional and social change for teenagers. There is an increased need for more

independence and control over their lives, and to fit in well with their friends. They also take on new responsibilities such as managing their health needs, self-infusions, haemophilia check-ups and school commitments. However, for many children, accurate diary keeping and self-infusion may start at an earlier age. Learning these skills can help increase a sense of control, independence and self-esteem. But at the same time, the desire to participate in all the same sports and physical activities as other teenagers presents many challenges and can lead to risky behaviour. Fitting in with friends and classmates becomes very important. Teenagers can be very sensitive about their body image. Some teenagers experience anger because they feel different and left out. Some reject haemophilia by ignoring their routine care. Teenagers may feel emotional stress. A child who has been encouraged to talk about his or her feelings, and taught how to take good care of his or her body, will likely be able to deal better with teenage challenges.

Transitioning from paediatric to adult services

The concept of moving on or transitioning from the familiar and trusted environment of the paediatric service at Childrens Health Ireland at Crumlin to the adult service at St. James's Hospital, can bring about mixed feelings for families. There may be feelings of anxiety around meeting a new team of health professionals, the logistics of finding your way around a different hospital and even finding a parking space! For others, there may be a sense of excitement and anticipation, in other words, a fresh start. This process will become geographically easier from 2023 when the new Children's hospital Ireland opens on the St. James's campus thereby bringing together comprehensive care for all ages on one campus.

What is Transition?

Transition is the process of planning, preparing and moving from children's health services to adult health services. In the case of most people with bleeding disorders living in Ireland, this involves a transfer of care from the paediatric team at CHI at Crumlin to the National Coagulation Centre (NCC) at St James's Hospital.

The transition process actually starts at the time of diagnosis and continues throughout childhood as children become more independent and begin to take ownership of the management of their bleeding disorder. Examples of this ongoing process can include; starting pre-school, encouraging a 10-year-old to self-infuse and encouraging a 15-year-old to have a discussion with their doctor on their own for part of a clinic visit.

While transition is an ongoing process that begins in early childhood, many of the big steps occur during the teenage years. Most young people move over to the NCC between the ages of 16 and 18 years. The health care professionals involved in the transition process are aware that this is often a period of significant change, whereby physical development and growth can be accompanied by many new emotional and social challenges.

The transition process may progress differently for each individual and family, for example, it may take into account if a young person is undertaking state exams or siblings close in age who would like to transition together. While there are guidelines in place, the transition process can be flexible and tailored to meet the individual patient's needs.

What to expect at the Transition Clinic?

The Transition Clinic is a designated outpatient clinic for young people at the National Coagulation Centre (NCC). Prior to the first Transition Clinic appointment at St James's Hospital, the team at CHI at Crumlin will have forwarded a transfer summary to the team at the NCC. This is useful for two reasons, it helps to plan clinics and it also gives us an idea of what health care professionals the young person may need to see on their first visit.

Initially, the young person will meet a doctor, a nurse and a social worker. Depending on their needs and concerns they may meet other members of the team such as the dentist, physiotherapist or psychologist. At the first visit to a Transition Clinic, the young person will be given a tour of the centre and introduced to team members.

An important part of the process is meeting and getting to know the team who will support the young person in the adult service. Meeting new health care professionals and building a relationship of trust will take some time. Transition can be daunting and it is important to remember that the health professionals in both hospitals are there to support and guide young people and their parents throughout the process. This is a team effort that includes the young person, parents and healthcare providers.

Health care professionals at the NCC focus on adult health needs and medical issues, the team works directly with the young person. From the first Transition Clinic appointment, the young person will continue to be supported to take responsibility for their own health needs. While the young person will have the option of attending their appointment on their own or with a family member, they will be encouraged to speak on their own behalf and to meet with health care professionals on their own.

The young person will also be provided with relevant patient booklets along with the contact details of the NCC and that of the out of hours service. Return appointments are made at the desk before leaving. All patients receive a reminder of their appointment by text message if a mobile number has been provided. Normally the young person attends two Transition Clinics before attending regular adult clinics.

In conclusion

The transition to adulthood and adult services involves major changes for all involved. However, it is important to remember that moving on to an adult treatment centre, is one of the final steps towards independence and empowering young people in the management of their own bleeding disorder. It is a crucial step as they embark on their exciting journey into adulthood.

Fear of needles/ procedural distress

Nobody likes getting injections and a fear of needles and painful procedures in children is normal. Thankfully needle phobia is uncommon and an inappropriate term for the normal distress and anxiety experienced by children and young people when they have needle procedures. Fear is a normal response to a threat, (in this case, needle insertion), that involves three response systems, physiological arousal, covert feelings and thoughts and overt behavioural expressions. A phobia is considered to be an unreasonable response to unharmful stimuli. Clearly needles are harmful in that they physically hurt, so your child responding with fear is normal.

Helping your child cope:

It is helpful for parents to take as active a role as possible in supporting and encouraging their child through the needle insertion.

- Stay calm and supportive but firm and matter of fact.
- Try to involve your child even in small ways.
- Allow your child to have some choice where possible, e.g. which parent to sit with, or which arm to use (if possible). This will give them some sense of control.
- It is helpful to permit your child to be upset and not expect them to "be brave".
- Distraction – encourage your child to focus on something which will absorb his attention during the procedure
- Slow breathing – teach your child to breath in deeply and blow out the scary feelings.
- Afterwards always acknowledge and reward your child's efforts to comply with the needle procedure, even if it doesn't go particularly well.

While it is common for children to be afraid of needles, most will adjust and learn to cope with needles in their lives. If you or your child develop difficulties coping with needle procedures, it may be helpful to have a consultation with your medical teams' paediatric psychologist in order to develop an individualised age appropriate coping plan for your child. This may help prevent difficulties in the long term.
Paediatric psychologists will tailor the approach recommended to your child's age. With younger children they may use and

recommend medical play and distraction , while with older children and teenagers they often use cognitive-behavioural therapy (CBT) interventions. These are a group of procedures aimed at identifying unhelpful thoughts, attitudes and problem behaviour. The key idea in CBT is the idea that challenging our negative thinking can be helpful. Thoughts affect feelings and if we change our thoughts we may be able to change our feelings. CBT has been found to be helpful when used in combination with progressive muscle relaxation training, guided imagery and distraction. These approaches has been found effective in reducing stress related to needle-related procedural pain.

Veincare

A child with a bleeding disorder is well educated to understand the important role their veins have in their body, as they get infusions of factor.

Veins are blood vessels that carry blood around the body. But for people with bleeding disorders, they are much more; they are lifelines that literally carry factor around the body. Most children will have a port-a-cath inserted until their veins are strong enough to take regular infusions. However, once a vein becomes strong there is no guarantee it will stay strong forever. Veins, like most tools, can become damaged because of overuse, so it is important to take care of your veins as you grow older – it is never too late to start.

Some examples of how to maintain good veins include:

Vein Rotation

Find three of four veins that are easy to access and rotate between them when administering factor. If you have time to take your factor, use the more difficult vein. Never rush when giving an infusion. You may find you are much quicker when using the easy or "reliable" vein, but if you overuse this vein it won't be reliable for too long.

Arm Exercises

Keeping fit and healthy helps strengthen muscles and joints which can help develop good veins.

Vein Exercises

The staff at the haemophilia treatment centre can help with some exercises to help build veins, but you can try the following:

- Secure a tourniquet to your arm and squeeze a fist or a rubber ball in your hand. Repeat this twice a day every day for no longer than 5 minutes at a time.

Secret Tips!

If you are in hospital for infusions or blood tests, ask the nurse/doctor not to use your "reliable" vein. Staff are trained to access more difficult veins and this will provide some rest for your reliable vein.

It is important to know what not to do:

- Do not infuse into an area that is swollen, inflamed or sore to touch.
- Do not put pressure on the injection site until the needle has been removed.

As we grow older we begin to see changes in our body. Our skin can wrinkle and sag, our hands become less steady and our eyesight can deteriorate. All of these make infusing more difficult. Therefore, the importance of good vein care is vital. So don't wait until it's too late, take action today and take care of your veins.

Dental Care

Taking oral health seriously

It is important for children with bleeding disorders to take good care of their teeth and gums. Regular visits to the dentist will reduce the chance of future problems such as extractions or mouth infections, which can lead to further problems.

Attending the Dentist

Your child can attend their own dentist for routine care such as check-ups, x-rays, fissure sealants, fluoride treatments, fillings, cleanings, root canal treatments and crowns. You should inform the dentist about the bleeding disorder and advise them that further information is available from your child's haematology treatment centre. Should your child need a tooth extraction or certain injections, your dentist should contact your haematology consultant or nurse as these types of treatment may need to be provided at a specialist centre. Your dentist will advise you and your child on how best to avoid dental problems and will provide regular interventions such as fluoride treatments and fissure sealants to prevent dental disease. Every child with a bleeding disorder should attend their dentist twice a year.

Top tips for healthy mouths

- Use a soft, child sized toothbrush to brush teeth twice a day.
- Children over two years should have their teeth brushed with a full strength fluoridated toothpaste.
- For children under two, you should consult their dentist regarding toothpaste use.
- Drink only milk and water during the day.
- Drink only water at night.
- Avoid fizzy drinks, juice and diluted fruit drinks.
- Restrict sweets and sugary foods.
- Attend the dentist by one year of age.
- Have a check-up every six months.

Siblings

When one person in a family has a bleeding disorder, it can be very easy to ignore some of the needs of other family members. This can lead to jealousy and resentment, particularly from siblings. On the other hand, it is also easy to leave all the responsibility for looking after a boy with haemophilia to one member of the family, often his mother.

As far as possible, try to involve the whole family in all activities. In this way, not only will everyone carry the responsibility for your child with haemophilia, but everyone is also expected to take an interest in whatever anyone in the family is doing. Asking your other children to help care for their sister or brother can help them come to terms with his condition and feel 'important'. Making sure that he participates in all the family's activities will help to switch attention away from him and towards the other individuals and the family as a whole.

At some time, everyone in the family will feel the pressure of having a brother or son with haemophilia. This is only to be expected. However, with a little compromise and understanding from everyone, these problems can and will be overcome.

School

What should teachers look out for?

Many incidents that you might expect to cause problems can easily be dealt with by simple first aid. Others cannot. These are the ones that cause a bleed.

Cuts, grazes and bruises

The minor cuts, grazes and bruises that are an everyday part of childhood do not normally cause any significantly greater problem for boys with haemophilia than for any other and can be dealt with by normal first aid measures.

Point of note: People with haemophilia do not have thinner blood and do not bleed any faster than anyone else – nose bleeds, cuts and grazes will slow and stop with standard, basic first aid treatments.

Obviously, when dealing with any form of open bleeding or blood spillage, all the normal precautions should be observed (e.g. wearing gloves, cleaning up with dilute bleach solution and paper towels, safe disposal in a sealed polythene bag).

Cuts and grazes should be covered with a plaster and/or bandage and be given direct pressure for a few minutes. Deep cuts, of the sort that might need stitching, should be covered and then referred to your local Haemophilia Centre for treatment.

Bruises are only a problem if they are particularly painful, which may be an indication of a deeper underlying bleed, or if they are the result of a head injury, which is always a cause for concern. Injuries to the scrotum should also always be taken seriously and referred for treatment.

Never give any medicine containing aspirin (acetylsalicylic acid) to a person with a bleeding disorder. Aspirin slows clotting and will make the problem worse.

Nose Bleeds

These may normally be stopped by applying firm pressure to the affected nostril for 10-20 minutes, or with an ice pack applied to the bride of the nose for a maximum of 5 minutes, while the boy is kept sitting upright. If these measures do not succeed, he should be referred back to his parents or to his Haemophilia Centre.

Bleeding from the tongue, or in the mouth

Any bleeding with the mouth is harder to deal with because any clots that form tend to be either dislodged by the tongue or food or washed away by the saliva. Sucking an ice cube may work, but usually bleeding inside the mouth will need treating at the Haemophilia Centre.

Head injuries

Any head injury may potentially be serious, since there is always the possibility of bleeding within the skull, which would put pressure on the brain. Therefore, even if you judge that the 'injury' really is trivial, you should still keep a close eye on the child for the rest of the day and tell the parents, so that you, or they, do not miss any of the signs that the injury may be more serious than it at first appeared.

Bumps

Children often bump their heads. If the boy is not distressed and not in pain, the bump is unlikely to have caused a bleed and probably does not need treating. Nevertheless, watch him carefully and, if in doubt, get help.

Minor injuries

Head injuries that cause bruising or involve even small cuts should always be treated by the parents or the Haemophilia Centre.

Serious injuries

These are usually the result of a hard blow to the head. Any injury which knocks the boy unconscious is always serious. Remember, any head injury may be serious, even if at first sight you might not expect it to be.

If a boy shows any of these signs, whatever his injury, he needs treatment as soon as possible, so contact his parent and or the haemophilia treatment centre.

- Persistent or worsening headache
- Nausea and/or vomiting
- Drowsiness or abnormal behaviour
- Weakness in one or more limbs
- Clumsiness or poor concentration
- Neck stiffness or pain
- Blurred or double vision
- Going cross eyed
- Loss of balance
- Fits or convulsions

Joint bleeds

Older boys should be able to tell you themselves if they are having a bleed. Younger ones may indicate that there is a problem by appearing upset or by 'protecting' a limb by limping or not using it. The commonest sites for joint bleeds are: Shoulder, Elbow, Wrist, Hip, Knee, Ankle and the main signs are:

- Pain or a 'funny feeling'
- Swelling of the affected joint or muscle
- Heat over the affected joint or muscle
- Loss or impairment of movement

School Related Issues

Some children have more problems adjusting to the classroom environment than others, and some children experience more difficulty learning certain subjects. These children are often the target of pranks and are frequently very unhappy at school. Imagine yourself doing a job every day where you are criticised, blamed and cannot understand how to do your work. You are paid for the amount of work you do. But regardless of your difficulty with the tasks of the job, you are still expected to perform as well as others, and to earn as much money. How would you feel? You would probably quit. When children have problems at school, they cannot quit. Sometimes they do the next big thing by becoming the class clown, so no one expects them to perform, or becoming aggressive so they get to spend most of their time in detention where they cannot do their work. This leads to problems at home as well because most parents feel they must do something about objectionable school behaviour. So, school-related problems result in problems both at school and at home.

Children with bleeding disorders may have to go to school one or two days on crutches, be absent a day or two, or appear with an ace wrap on their elbow. Some children have been infused with factor at school by one of their parents.

When the child's difficulties at school remain unrecognised and/or untreated, things get worse. School personnel blame the parents, and parents blame the schools. Rather than letting this occur, the best thing to do is meet with the teacher and other school professionals to find out exactly what is going on. When parents are involved and demonstrate that they are following through with their child, the blaming cycle often stops and the business of obtaining the best educational planning for their child can be accomplished. If your child seems to have any of these problems, you can do a lot for him by getting the recommended evaluations and accepting that some things in school may be harder for your child than for others. The most important issue here is understanding that your child needs help from you and the school.

Sports

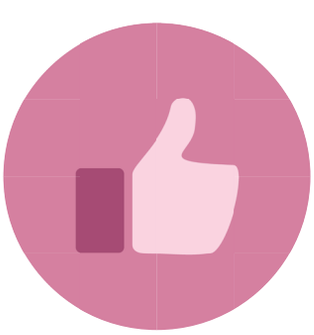
Yes, you can play sports!

Exercise is very good for you and your family, and it is very beneficial. Most sporting activities are safe, but there are some sports such as boxing and rugby which are not recommended for children with haemophilia. Just because you have haemophilia does not mean you cannot play sports! If you keep yourself fit and play sports you are less likely to get a bleed. Everyone can enjoy sports no matter where you live or what age you are. Swimming is great fun and fantastic exercise for children with haemophilia. Cycling, golf, tennis, athletics and fishing are also recommended.

The more you look after yourself and keep fit the more this will help your muscles, and you will feel much healthier. Why don't you get your whole family involved in a sport? Now I know some of you would just love to play rugby or American football or even wrestling, but just think about it, it would not be very nice to get a head injury would it? So you have to think about it very hard before you pick a sport.

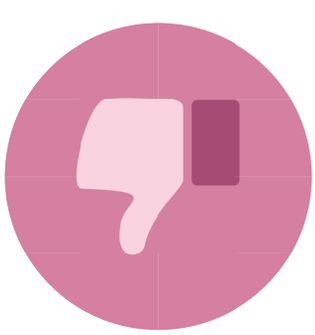
Sports recommended for children with haemophilia

Cycling
Tennis
Athletics
Fishing
Frisbee
Hiking
Swimming
Tai Chi
Walking
Golf



Sports not recommended for children with haemophilia

Baseball
Diving: Recreational
Karate
Mountain Biking
Skateboarding
River Rafting
Rock Climbing
Roller Blading / Skating
Rowing
Skiing
Snowboarding
Hurling
Gaelic Football
Diving: Competitive
Rugby
Wrestling



Travelling

Checklist:

- Treatment
- Cooler bag
- EHIC Card
- Passport
- Visa if necessary
- Other regular medications
- Letter for airport security / customs
- Insurance documents

Before you travel

- For travelling within Europe you should have an EHIC card (European Health Insurance Card). This entitles you to necessary healthcare in the public system of any EU / EEA member state, or Switzerland if you become ill or injured while visiting another country. You can apply for the card online at www.ehic.ie
- Visa requirements should be checked well in advance of travel.
- Medical insurance should be checked well in advance of departure as travel insurance can be difficult to obtain for a person with haemophilia. A company which does provide insurance cover for a pre existing medical condition including haemophilia is Blue Insurance. They can be contacted on 0818 444 449 or at www.blueinsurance.ie. Full disclosure is required and there will be some loading however this is not a significant amount.
- You will require a letter from your treatment centre for airport security/customs. The letter should be on headed paper and confirm that you are carrying treatment, needles, syringes etc, as well as relevant information on any metal implants (such as knees or hips) that may set off the security scanners. The letter should not be more than six months old and should be updated as necessary. This letter should be signed by a consultant doctor. If travelling to the USA, the letter should ideally specify the brand name of the factor you will be carrying. To allow for some delays you should pass through security as early as possible.
- Always carry your own treatment, syringes, needles etc. with you as products vary in different countries. A general rule when packing factor, you should bring your regular dose for the duration of your trip plus sufficient factor for at least one extra 100% rise.
- If flying, always carry the factor concentrates and infusion supplies required in your hand luggage. Never place them in checked in luggage. There is no guarantee that your luggage will arrive at your destination.

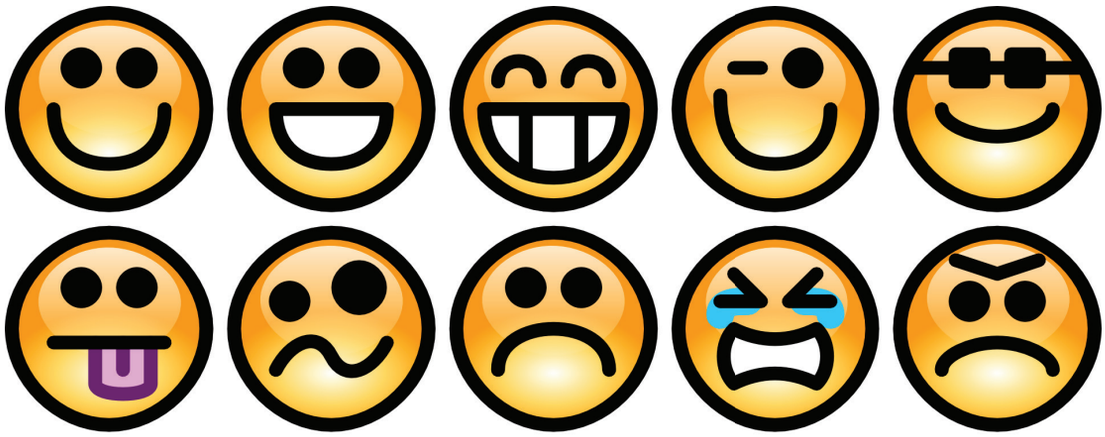
- It may be useful to pack one medication information insert and one box cover. In the event that you cannot treat yourself, or if there is a language barrier, this would be useful for the treating doctor.
- National haemophilia organisations and treatment centres can also be a source of assistance if needed while travelling. You will find these listed in the WFH Passport which is available online at wfh.org or on haemophilia.ie. Alternatively you can contact us at any stage for information on the country you are visiting. Having this information in advance allows you to plan in case of an emergency as not every hospital will have a treatment centre.
- Take a photo of your passport photo page and crucial travel documents and keep on your phone. You could also photocopy all necessary travel documents (tickets, passport, visa, one form of ID, travel letters, insurance policy, etc.) and leave them in an envelope at home where they can be accessed easily in case any of these documents get lost and need to be replaced. A photocopy will speed up the process of replacment documents. Another option would be to place an extra set of copies in your checked in luggage, or you could scan travel documents and send them to yourself in an email.
- If you are planning a long haul flight it may be useful to take some treatment before you leave.

While travelling

- Ensure you have your Bleeding Disorder Alert Card in your wallet. Bring an I.H.S. travel card which provides useful phrases in several languages.
- Bring details of any insurance cover policy numbers, and contact details with you.
- When in transit (bus, train or plane) always carry your treatment in your hand luggage to prevent loss or breakages. If the trip is long and you have a lot of treatment, carry all the treatment and possibly 5-6 injection kits in your hand luggage. The rest of the injection kits can be stored in your main luggage. You can leave your factor in your carry on bag. However, it is recommended to put your medication (in cooler bag or appropriate bag) in the tray with your coins, watch, wallet etc. when going through security at the airport. You may be asked to open the bag but if you have your letter you should not have any problem.
- It is important to know how long it is recommended that treatment can be stored out of the refrigerator and also the temperature range at which it can be stored outside of the refrigerator.
- It may also be useful to know if your hotel or hostel has a secure refrigerator, where you can store your treatment, especially in warmer countries.

Emergencies

It is highly recommended that you should have a letter on headed paper from your treatment centre with information about your bleeding disorder, the usual treatment you receive and any other medical conditions or information that may be useful in case of emergency. This letter should not be more than six months old. If you need to attend a hospital while you are away, if possible treat yourself first. Accident and Emergency rooms can be slow at the best of times and if a translator is required this may add to the delay in getting your treatment.



Coping

Feelings

After the initial diagnosis of your child's condition there is usually a period of heightened stress and upheaval in the family. As stated earlier it is common to experience a range of strong emotions, just as one would after a loss or bereavement. Many people say this grief reaction is as a result of the loss of a "healthy child". You may experience, shock, denial, sadness, anger, anxiety, guilt and finally acceptance. This process does not happen in a neat order or sequence. It is important to pay attention to your self-care and get support so that you can learn to cope with the new challenges and demands of having a child with haemophilia.

We all have a whole range of different ways we cope with challenges / stress in life. We can learn from what we have found helpful in the past. Everybody copes differently, the following are some coping strategies for you to consider:

Information – get accurate information from reliable sources. Information gives us a greater sense of confidence so that we can predict and plan better.

Meeting others - in a similar situation.

Social support – your partner, best friend, family member, work colleague. Reach out and reduce your isolation. Accept offers of emotional support and practical help.

Talking and relationships – It might sound obvious but just talking out loud about your worries with someone can give you a distance from the worries and help you come up with ideas about how to deal with your problems. Often couples stop talking to each other after a diagnosis because the are worried about burdening their partner with their worries and as a result you can end up even more isolated. Make a conscious effort to regularly talk through your worries and stay connected.

Relaxation and Self-care – at times of stress we often neglect our own physical and mental health. Try to eat well and get regular physical exercise. Keep an eye on your caffeine and alcohol intake. Think about what helps you de-stress and relax, e.g., reading, walkng, listening to music, yoga, whatever it is try to make room in your week for yourself.

Learn to relax – the physical symptoms of anxiety and stress occur because adrenaline is released by the nervous system into the blood stream and affects organs such as the heart, stomach and muscles. Relaxation and breathing exercises can help you control these symptoms.

Understand that stress is a normal human response in the face of too many demands. Be patient with yourself and your child.

Shared responsibility – between the family, with the child and with your medical team. Not expecting perfection.

Temperament, personality and physical capacity

People have some characteristics that are similar overall, and some that are different. Anyone who has more than one child will be able to say that each one has individual differences as well. Some children respond more readily to verbal commands than others. Other children may be more impulsive, active, obstinate, moody or social.

Recognising that differences in temperament with children in the same family is a natural phenomenon. It helps parents to understand that these factors are not intentional on the part of the child, nor are they a result of how the child had been parented. Some children have a "different temperament", which means that they may be easily distracted, overactive, or may react more intensely to environmental stimuli like sound and touch. Understanding this often makes it easier to be objective when dealing with childish offenses. Remember that your child's personality will be the result of factors within the child and events that occur during his developmental years - his unique qualities, environment, and your style of parenting.

It is also important that a parent understands each of their children's unique physical conditions and capabilities to help him/her function optimally.

Behavioural Issues

When a young child with a bleeding disorder has a tantrum it can be especially upsetting for parents. There is real cause for concern because injury can result in a bleed. Parents are wise to refuse to let their child become domineering by giving into inappropriate behaviour. We recommend that parents move the child to a safe place and then use their ignoring skills. Safe places could be such areas as a sofa or a carpeted area away from sharp corners and hard surfaces. If tantrums are a problem pad an area with pillows in advance. Then when the tantrum occurs take the child to the area. You may say something like "OK, here is the spot where you can do this. When you're finished you can come back and play." Do not make the mistake of standing by and watching noticeably or the child will perceive this as attention and increase the tantrum. Walk away and watch out of the corner of your eye. Then when it is over, you might invite the child to join you in some activity but do not mention the tantrum or offer any comfort. Children need to learn to deal with frustration. When the tantrum is over, the child may be emotionally drained, but he has conquered the frustration. Let him experience his own sense of control over his emotions. This is not a good time to offer comfort and sympathy. This is one time when you can take it for granted.

Support

The Irish Haemophilia Society offers outreach and support services including home and hospital visits, regional visits, school visits and counselling to:

- Children with haemophilia and their families
- Families with a new diagnosis of haemophilia
- Carriers of haemophilia
- Persons with von Willebrand's Disorder
- Women with bleeding disorders
- Individuals with rare bleeding disorders

The ongoing level of personal contact between the I.H.S. staff and I.H.S. members is very high. Each member of staff is trained and educated sufficiently about haemophilia and the ramifications of living with a bleeding disorder to enable them to proactively engage with and assist members. Members build relationships with the staff team from their attendance at conferences and meetings. These relationships are very helpful in allowing us to optimally help the members.

Meeting members in their homes allows us to integrate our understanding of the issues which specifically affect members in that area and allow us to optimise our advocacy efforts on their behalf.

Being in hospital can be a scary and lonely time. The I.H.S. office is located in Dublin City Centre so it is easy for staff to attend Children's Health Ireland at Crumlin. If you or your child is in hospital and you feel like you need support, a chat or maybe a newspaper don't hesitate to contact the office. Remember, the staff are here for you! We can offer you assistance and practical support during your hospital stay.

If your child is starting crèche, playschool or primary school and you are a little worried, please call the office on 01 6579900. We can arrange to visit the school and will talk to the teachers and educate them about haemophilia or a related bleeding disorder. We can also supply the school with I.H.S. publications. This service can be arranged at the school's convenience.

Planning Your Family

Genetic testing

There are a number of genetic tests that can be done to determine carrier status. These tests are complex. Results are more reliable if DNA from a family member with haemophilia is also available. In Ireland, the policy is that a possible carrier must be old enough to understand the implications of the test results before genetic testing is carried out. This is usually after 16 years of age. The sooner a girl is tested after this age, the more time she has to come to terms with being a carrier. It also prepares her for eventual decisions about pregnancy and childbirth. There are some arguments against carrier testing by genetic analysis at a young age. It may affect the child's self-esteem or the family's perception of the child. Some families feel the only reason to know about carrier status is for family planning. If a girl is an obligatory carrier (daughter of a man with haemophilia), her factor level should be checked at the earliest possible age in case she has a low factor level. This also applies to potential carriers. It is important to know the girl's factor level in case of trauma or requirement for surgery

Psychosocial issues

People cope in different ways when they are faced with new information. This depends on age, reasons for seeking the test and on any previous experience they might have with the condition. Carriers can receive this diagnosis at a young age due to health problems, or as adults after the birth of a child with haemophilia in the family. A girl or woman can experience a wide range of emotions when she finds out that she's a carrier. She may be afraid of what it will mean for her, her relationships and any children she may have. She may feel a sense of loss that she is different from everyone else and may even fall into a pattern of denial and refuse to acknowledge the diagnosis. When a son is diagnosed with haemophilia and there is no family history, parents also have to deal with the possibility that his sisters are carriers. Parents may feel like this is a second blow. A person's decision to progress with carrier testing can affect the extended family. Genetic counsellors can provide suggestions on how to inform other at-risk female relatives such as sisters, daughters or nieces.

Where?

Call the National Coagulation Centre on 01 4162141 / 01 4162142 for advice and an appointment.

Please also note:

- It will speed up the diagnosis if a member of your family with or carrying haemophilia has already had their genetic mutation identified as the mutation, if present, will be the same for any carrier or person with haemophilia in the same family

- The Clinic is co-ordinated by a Clinical Nurse Specialist
- A follow up appointment is sent when results are available
- A confirmatory sample is requested when attending for results of initial sample
- Results may take some time especially if the genetic mutation has not been previously identified in your family

Pregnancy in women with bleeding disorders

When a woman with a bleeding disorder 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. It is important that prospective mothers bring copies of their delivery plan to the hospital when they go in to have the baby.

Remember

If there is a family history of haemophilia, ensure that you are aware of your carrier status before pregnancy occurs. Inform your obstetrician of your bleeding disorder early in your pregnancy.

Appendix 1

Port-a-caths / PICC Lines

What is a port?

A port is a special device, which is used to allow easier, more dependable access to your child's veins. It is made up of an injection port and a long hollow tube. The injection port is attached to one end of the tube and sits underneath the skin on your child's chest. The other end of the tube is placed in a vein close to your child's heart. When your child needs an injection, a special needle is inserted through the skin into the injection port using a sterile procedure allowing access to his/her veins.

When you and the haemophilia team have decided your child needs a port, he/she will be referred to the surgeon who will send you an appointment to come to the surgical clinic to discuss the procedure.
The surgeon will explain the procedure, the alternatives and the risks involved and answer any questions you may have. A date for the procedure will be arranged and you will receive notification of this date from the Admissions Department.

It will be necessary to bring your child to your haemophilia centre the week before surgery to have a blood test performed to check for inhibitors. The haemophilia nurse will organise this.

How is a port inserted?

Your child will go to the theatre and have a general anaesthetic for the operation. He/she will be in the theatre for about two hours in total. The surgeon will insert the port. This usually takes about 30-60 minutes. The rest of the time is spent putting your child to sleep and waking him/her up again.

What happens when I take my child to the hospital to have a port?

Admission: For children with haemophilia who need a port have the procedure at Children's Hospital Ireland in Crumlin, a six-day stay in hospital is usually required. Your child will be admitted to the hospital the day before the operation. You will need to phone Admissions that morning to confirm there is a bed available and confirm the time of admission (usually 2 p.m.). On arrival to the hospital, you will attend the Admissions Office and from here you will be directed to the ward. Parents are welcome to stay either by the bedside or in parents' accommodation. Further information is available in the patient's handbook that will be provided by one of the haemophilia team.

Assessment: When you arrive on the ward a nurse will carry out a nursing assessment. A surgical doctor will examine your child.

Consent: The doctor will explain the procedure, answer any questions you may have and ask you to sign a form declaring that you have received sufficient information and you are giving your consent for the procedure. Do not sign this form unless you are happy you have all the information you need to fully understand the procedure. During the evening or night, a specialised nurse from the IV team will put in a cannula and take some bloods. Your child will need to fast for the operation. He/she will not be allowed food after 2 a.m. but may have a drink of clear fluids up until 6 a.m. the morning of the operation.

The morning of the operation

A dose of coagulation factor concentrate will be administered to your child to bring factor level up to normal. A blood sample must be obtained from your child post factor administration. Once this blood is taken your child will be attached to a drip, which will administer coagulation factor concentrate for the next five days to keep his/her levels normal. He/she will have regular blood tests to make sure the infusion is maintaining his/her factor levels. As soon as the factor levels are confirmed your child will be taken to the operating theatre. All children with haemophilia having a port inserted will be in the theatre department before midday. One parent may go to the theatre and stay with the child until he/she is asleep.

After the operation?

After the operation, your child will be sent back to the recovery room until he/she is awake and ready to go back to the ward. A nurse from the ward will collect your child from the recovery room. You may go to the recovery room with the nurse. Your child may be irritable when he/she first awakens. This may be due to the anaesthetic or because he/she is sore or hungry. There will be a bandage over the wound to prevent bleeding and infection. This bandage should not be removed for 48 hours. There will usually, but not always be a needle in the port. If there is there will be a short tube with a bung on the end coming from the port. If there is a needle in the port, it may be used immediately if absolutely necessary. Factor level cannot be taken from the port.

On return to the ward, the nurses will closely monitor your child, his/her pulse and blood pressure, the wound and the coagulation factor infusion. Your child will receive pain relief medication during the procedure and should continue to have pain relief regularly for 24 – 48 hours afterwards. Paracetamol is usually sufficient, but codeine may be required. It is important that your child has sufficient pain relief so he/she can move normally and not become stiff. As soon as your child is awake and alert, he/she can have something to eat and drink. It is a good idea to start with clear fluids and a light diet in case he/she is sick after the anaesthetic. As soon as they are tolerating food they can eat normally.

The factor infusion will continue for five days after the surgery to ensure your child's factor levels are normal until the wound is healed. This means he/she will be continuously attached to a drip, which will restrict his/her mobility for five days. Please bring toys or books to help amuse your child during this period.

The wound is generally closed with a dissolvable continuous stitch with steri-strips (plaster stitches) on the outside for added strength. The steri-strips should be left in place and allowed to fall off of their own accord. This may take 10 days.

What is a PICC Line / Broviac?

A PICC is an acronym for a Peripherally Inserted Central Catheter, and it is, in essence, a long IV line. It is usually inserted in a smaller vein in the upper arm (peripheral) and terminates in a larger vein in the chest near the heart (central). "Catheter" is just another name for "tube" in the medical lexicon.

PICC lines are inserted by certified PICC specialists, who use ultrasound and X-ray images to ensure that the line is in the correct spot. PICC lines can have 1-3 lumens, which allow medications to be attached and flow through the line. Additionally, blood draws for lab purposes can also be extracted from the line. The one pictured has two lumens, which have orange "caps" to help keep the line sterile. Sometimes these caps are blue as well.

A PICC line is a safe, stable, and effective way to deliver IV medications. They can stay in the body for weeks or months, alleviating the need to subject your veins to the numerous needle sticks necessary if the PICC was not there. Additionally, some medications are caustic to the vasculature; PICC lines allow these medications to be delivered to larger vessels that are less likely to be damaged. PICC lines are thus a very versatile choice for patients requiring long-term venous access for conditions.

Infection is always a risk with intravenous access but reported rates of PICC line infections are low if proper maintenance protocols are followed. The lines need to be flushed at regular intervals, and the dressing needs to be changed weekly.

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