Irish Haemophilia Society

Information for Teachers
&
Playgroup Leaders

Contents

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

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

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Introduction

Academically, there is no difference between a child with haemophilia or related bleeding disorder and a child with no bleeding disorder. However, sometimes children may miss school when they are recovering from a bleed. Haemophilia or a related bleeding disorder should not interfere with a child's education. Education is vitally important in today's world as it provides the child with every opportunity to participate fully in the activities of normal life.

Haemophilia is rare, so teachers and playgroup leaders may not be familiar with the condition. It is therefore very important that teachers, playgroup leaders, schools and creches are fully informed of the nature of haemophilia. Most severely affected children receive treatment at home, thus preventing most bleeding episodes.

If your child is starting creche, playschool or primary school and you are a little worried, we are here to help. 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 publications.

What is haemophilia?

When a person's blood does not clot properly the condition is known as haemophilia. A person with haemophilia will not bleed any faster than anyone else following an injury, but they can bleed for longer. In 70% of all cases, there is a family history of haemophilia. In as many as 30% of cases, the mother may not be aware that she is a carrier, or the condition may have occurred spontaneously. The two most common types of haemophilia are haemophilia A and haemophilia B. People with haemophilia A have reduced levels of a protein called factor eight (written as VIII). The people with haemophilia B have reduced levels of a protein called factor nine (IX).

Haemophilia Categories		
Mild Haemophilia	Moderate Haemophilia	Severe Haemophilia
Factor levels @ 5% - 40%	Factor levels @ 1% - 5%	Factor levels @ - 1%

Once established, the severity of haemophilia does not usually change during a person's lifetime and people in the same family normally inherit the same level of severity of haemophilia.

Can you explain the Clotting Mechanism?

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

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

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

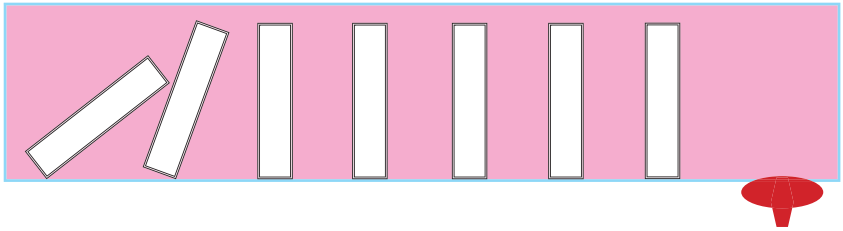
How will haemophilia affect a child?

Children with mild or moderate haemophilia do not usually have bleeding problems in everyday life. They tend only to have problems after an injury, an operation or dental treatment. Those with moderate haemophilia should nevertheless be aware that bleeds could happen from less severe injuries and that nosebleeds may be more frequent and last for longer.

In contrast to those with mild or moderate haemophilia, people with severe haemophilia have spontaneous bleeds into joints and muscles even when they have not been injured. They need treatment to stop these bleeds or to prevent them.

Normal Clotting

1.



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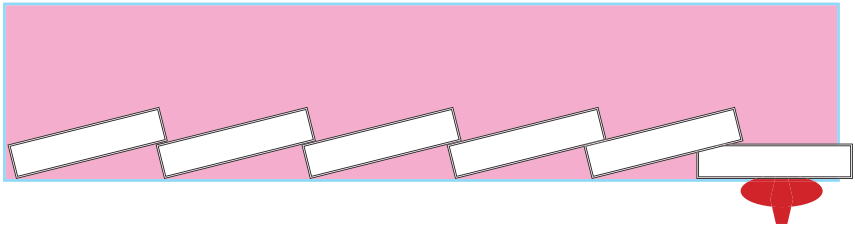
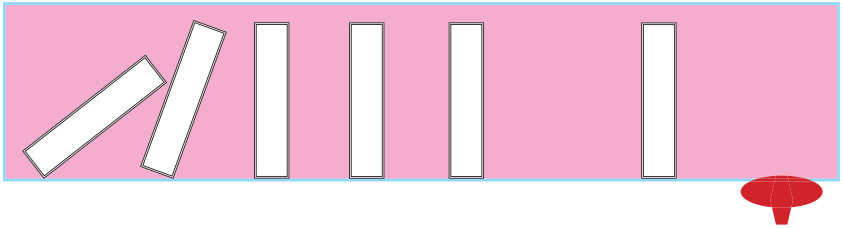


Haemophilia

1.



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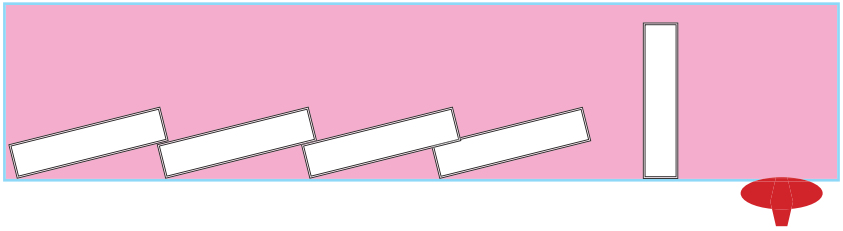
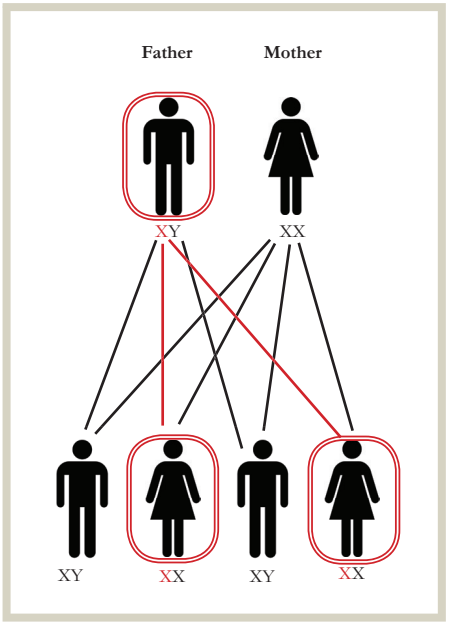


Can you explain the inheritance of haemophilia & vWD?

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

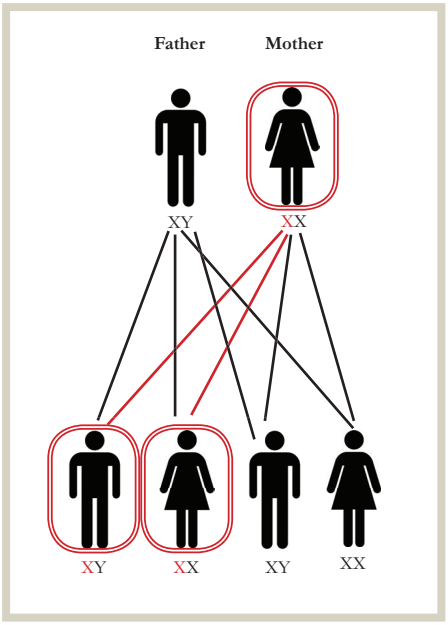
When the father has haemophilia and the mother is unaffected



None of the sons will have haemophilia.

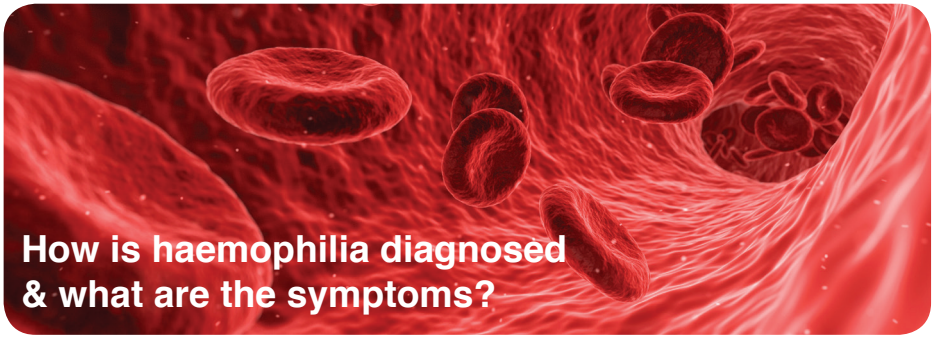
All of the daughters will carry the haemophilia gene

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



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

Von Willebrand's Disease (vWD) is slightly different. Both men and women have two von Willebrand factor genes. Therefore, men and women are equally affected by von Willebrand's disease. VWD is usually mild although it can be severe. Most people with vWD have the Type 1 or Type 2 form and will only experience a mild form of the condition and no medication will be required except when having surgery, and/or dental extractions. Rarely, an individual will have Type 3, which is severe vWD.



How is haemophilia diagnosed & what are the symptoms?

If doctors suspect haemophilia in a young child, there are simple lab tests that can be performed through a haemophilia treatment centre. A blood sample can be taken and tested to measure the amount of clotting factor activity in the blood. This will allow the doctor to determine if the person has haemophilia and the severity. Low levels of factor VIII indicate haemophilia A, whereas low levels of factor IX indicate haemophilia B.

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

The symptoms of haemophilia are as follows:

- big bruises
- bleeding into muscles and joints
- prolonged bleeding after getting a cut, removing a tooth or having surgery
- bleeding for a long time after an accident (especially after an injury to the head)

Also, keep an eye out for:

- the child being reluctant to move the affected limb/joint.
- the child complaining of pain in the affected joint/limb.
- swelling and heat in the affected joint or limb.

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

Can you explain how treatment works?

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. Some new preventative treatments can be given subcutaneously (under the skin) but treatment for a bleeding episode is always intravenous factor. Early treatment is the key to quick rehabilitation. Some parents have been trained to give treatment to their children. Some children self-infuse. Some children will need to be taken to the hospital for treatment. Bleeding can be internal or external.

Joint/muscle bleeds should be treated as soon as possible. If a child complains of chronic pain or swelling contact the child's parents and/or seek medical advice, elevate the limb and apply an ice pack and do not allow the child to use the limb until his parent/guardian arrives. Do not apply pressure until the child has had factor concentrate as constricting swelling can be dangerous. Children can sometimes return to school when recovering from a bleed. However, they should be allowed to elevate the affected limb and be excused from activities such as PE.

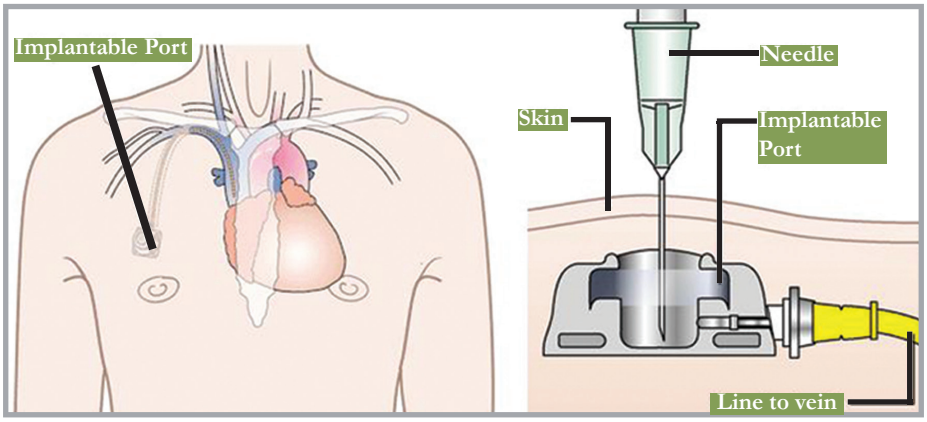
What is prophylaxis?

The word prophylaxis means a treatment that prevents something happening. For people with haemophilia it means treatment to prevent bleeding. Some people with haemophilia have factor added to their blood on a regular basis whether they have had a bleed or not. This type of treatment is called prophylaxis as the aim is to prevent bleeding. Regular prophylaxis injections at home can be with injections just under the skin (subcutaneous) or into a vein (intravenous), depending on the treatment of choice. This means that the factor will be in the blood ready to help

out if a bleed occurs. If the child is being treated with regular intravenous injections they may have a port-a-cath inserted.

What is a Port-a-cath?

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



Bleeding & Injuries

The most common types of bleeding in relation to haemophilia is internal bleeding into the joints or muscles and this bleeding 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.

Bleeds at a very young age usually occur as a result of a bang or fall. Superficial bruising is also a common form of bleed. Abrasions and small cuts cause no more trouble than in anyone else, they just need a sticky plaster. Joint bleeds are uncommon in the first three years of life.

Joints and Muscles

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. 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. Treatment for bleeding into the joints and muscles is coagulation factor concentrate (IV), followed by **PRICE**:

- Protection
- Rest
- Ice
- Compression
- Elevation

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 in 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.

Signs of a serious head injury

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

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

In the event of an emergency phone the 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.

Sports & Haemophilia

The first sport which most children enjoy is swimming. Swimming has been voted top sport for someone with haemophilia in an international survey of doctors by the World Federation of Hemophilia. Toddlers love going to the pool. Playing in warm water allows the exercise of all joints and muscle groups in the body without gravity. No child should grow into adolescence without being able to swim.

Appropriate sports and physical activities are generally recommended. Almost all sports and leisure activities are safe and they should play sports just like their friends do. Exercise will promote strong joints and muscles so they are less likely to bleed. It is important to encourage people with haemophilia or von Willebrand's to explore new physical challenges gradually and not to dash into potentially harmful activities. Only time will tell, if a particular activity is going to provoke unacceptable bleeding, and the chances of this happening will be reduced by careful attention to warm-ups, stretching exercises and cool-downs.

If it is found that a particular sport causes bleeds, it may be necessary to change to another sport or take treatment before taking part. Sports injuries occur just as much to people with a bleeding disorder as to anyone else. The treatment is the same.

For further information on sports, please refer to the Irish Haemophilia Society's booklet on sports & exercise, and activity table on pages 18 & 19.

Education & Haemophilia

Haemophilia should not interfere with a child's education. Education is vitally important in today's world as it provides the child with every opportunity to participate fully in activities of a normal life. Haemophilia is rare, so the school and teacher may not be familiar with the condition.

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

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

Creche & Playgroups

At playgroup, normal play activities present little in the way of problems and the child should be allowed to play alongside the other children. They may get some unsightly bruises occurring, but in general, these will not be a serious problem and no treatment will be necessary.

There is no reason why children with a bleeding disorder should not be allowed to use scissors and other sharp instruments. Like any child, they need to learn how to handle this equipment.

Primary School

Self-infusion is an important part of a person with haemophilia's life during primary school as it is the time when children start getting interested in learning how to treat themselves. Self-infusion is when a person with haemophilia gives treatment to themselves. Before starting self-infusion, a child with haemophilia is entirely dependent on someone else to infuse their factor. They are either unable to go on school trips or outings or one of their parents may need to go with them on every trip. This can lead to a lack of self-confidence or even frustration as they just want to be like their friends.

When a child starts to self-infuse, it is the next stage in them taking control of their haemophilia. They can now take care of themselves and that creates a feeling of independence and confidence. Self-infusing for the first time can bring fear which is quickly overwhelmed by pride and happiness. Starting self-infusion can be daunting for parents and children, especially when things are going well as they are.

In case you think your child is too young, the average age when children start to self-infuse is 7-9 years old, but there have been cases of younger children than this self-infusing.

Secondary School

Adolescents have the job of crossing that bridge as part of growing up. The parents have the role to decide when boundaries are necessary. Many new experiences must occur for the adolescent to cross that bridge. First is the need to have the physical abilities to drive the car. As the adolescent's body grows into puberty, so does the desire to get to the other side of that bridge to meet new people. The adolescent begins to develop the physical features of an adult. He is taller, developing and his body is stronger and more able to perform complex physical tasks. He is also developing intellectually and emotionally.

Some examples of these decisions are: future school and career choices, learning to drive, how to earn, use and save money, how to take care of personal hygiene, clothing, cooking, who to choose as friends, how to take care of his medical needs, how to get a date, who to date, how to plan a date, who and when to tell others about his haemophilia, etc. Each of these decisions is important in that they set patterns for how he will handle decision making as an adult. It is a time of experimentation, so mistakes are made and learning occurs from those mistakes.

For the adolescent with a bleeding disorder some of the additional issues of transition to adulthood will be:

- Managing self-infusion when away from home
- Maintaining infusion supplies
- Learning to consistently & promptly identify bleeds
- Completing infusion logs at the time of each infusion
- Making & keeping clinic appointments (call to reschedule when not attending)
- Doing home exercises as instructed
- Following what the doctors and nurses advise him to do

Some Points to Remember

- As a child grows, he should be allowed out to play with other children and eventually go to playschool. The advice that follows is what I give to all parents.

- All bumps or knocks to the head should be treated seriously. You should bring your child to your treatment centre immediately – do not wait for symptoms to appear.

- Do remember that any medication containing Aspirin should not be given to anyone with a bleeding disorder as this can cause bleeding. Always check with your doctor which medicine is safe.

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

- And remember your child is a normal child who just happens to have a bleeding disorder. Do not let him or his condition take over the family.

- There are some funny myths about haemophilia, and I think it also worthwhile reassuring teachers that the condition presents no threat to other children.

Table of Activity Ratings

Activity	Category
Aquatics	1
Archery	1
Badminton	1
Basketball	1.5 - 2
BMX Racing	3
Body Sculpt Class	1.5
Boot Camp Workout	2
Bowling	2
Boxing	3
Canoeing	1.5 - 2.5
Cardio Kickboxing Class	2
Circuit Training	1.5
Cycling	1.5 - 3
Dance	1.5 - 3
Diving Competitive	2.5 - 3
Diving Recreational	2
Elliptical Machine	1
Fishing	1.5
Football (American)	3
Football (Gaelic)	2.5
Frisbee	1 - 1.5
Golf	1
Gymnastics	2.5 - 3
Handball	2.5
Hiking	1 - 1.5
Hockey (Street / Field)	2.5 - 3
Horseback Riding	1.5 - 2
Hurling	3
Indoor Cycling	1.5 - 2
Jet-sking	2.5 - 3
Jumping Rope	2

Table of Activity Ratings

1	1.5	2	2.5	3
Low Risk	Low to Moderate Risk	Moderate Risk	Moderate to High Risk	High Risk

Activity	Category
Aquatics	1
Karting	2.5
Kayaking	1.5 - 2
Lacrosse	3
Tai Chi	1
Martial Arts	3
Motocross / Motorcycle	3
Mountain Biking	2.5
Pilates	1.5 - 2
Power Lifting	3
Rock Climbing (Indoor)	1.5 - 2
Rock Climbing	2.5 - 3
Rowing	1.5
Rugby	3
Running / Jogging	2
Scuba Diving	2 - 2.5
Skateboarding / Skating	1.5 - 2
Snowboarding	3
Soccer	2-3
Surfing	2 - 2.5
Swimming / Snorkeling	1
Tennis	2
Track & Field	2 - 2.5
Trampoline	2.5 - 3
Treadmill	1.5
Volleyball	2 - 2.5
Walking	1
Water Polo	2.5
Wrestling	3
Yoga	1.5 - 2

Personal Information For Teachers & Playgroup Leaders

Name of Child:	
Date of Birth:	
Address:	
Mother Home Phone:	Mobile:
Father Home Phone:	Mobile:
Emergency Contact:	Number:
Bleeding Disorder:	
Treatment:	
Haemophilia Treatment Centre:	
Name of consultant: Phone:	
Haemophilia Nurse Specialist: Phone:	
Special Requirements:	