

Haemophilia Information Day

5th September 2009

Cork University Hospital,
Wilton
Cork

The Child with Haemophilia

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*The Future is Brighter for Children with
Hereditary Bleeding Disorders*

What is Haemophilia?

- Haemophilia is an inherited bleeding disorder in which the blood fails to clot normally.
- People with haemophilia lack the normal levels of clotting factors.

The Child with Haemophilia

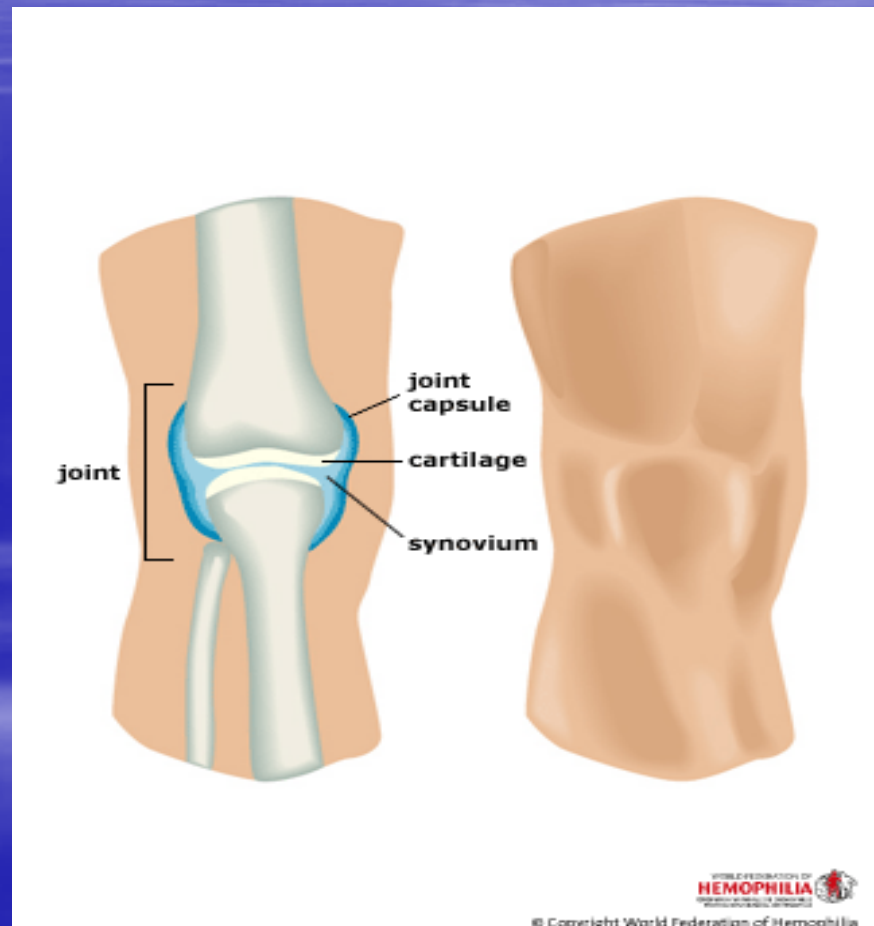
Is Haemophilia lifelong?

- A person born with haemophilia will have it for life.
- The level of factor VIII or IX in his blood usually stays the same throughout life.



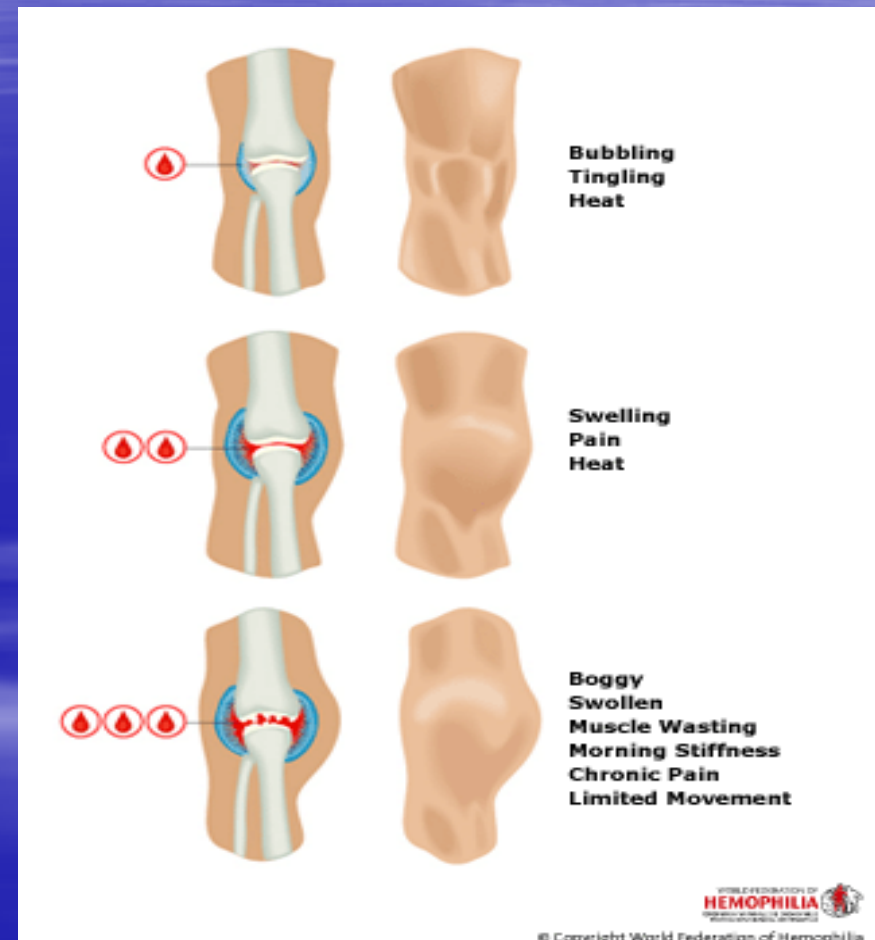
Assessing and Managing Bleeds

- **Joint** - The place where two bones meet. The ends of the bones are covered with a smooth surface called **cartilage**.
- The bones are partly held together by a **joint capsule**. The joint capsule has a lining called **synovium** with many capillaries (small blood vessels). It makes a slippery, oily fluid that helps the joint move easily.
- If the capillaries in the synovium are injured, they bleed. Often there is no clear reason for the bleed, esp. severe. In a person who does not have haemophilia, the clotting system stops the bleeding quickly. But in haemophilia, the bleeding continues. This causes the joint to swell and become painful.



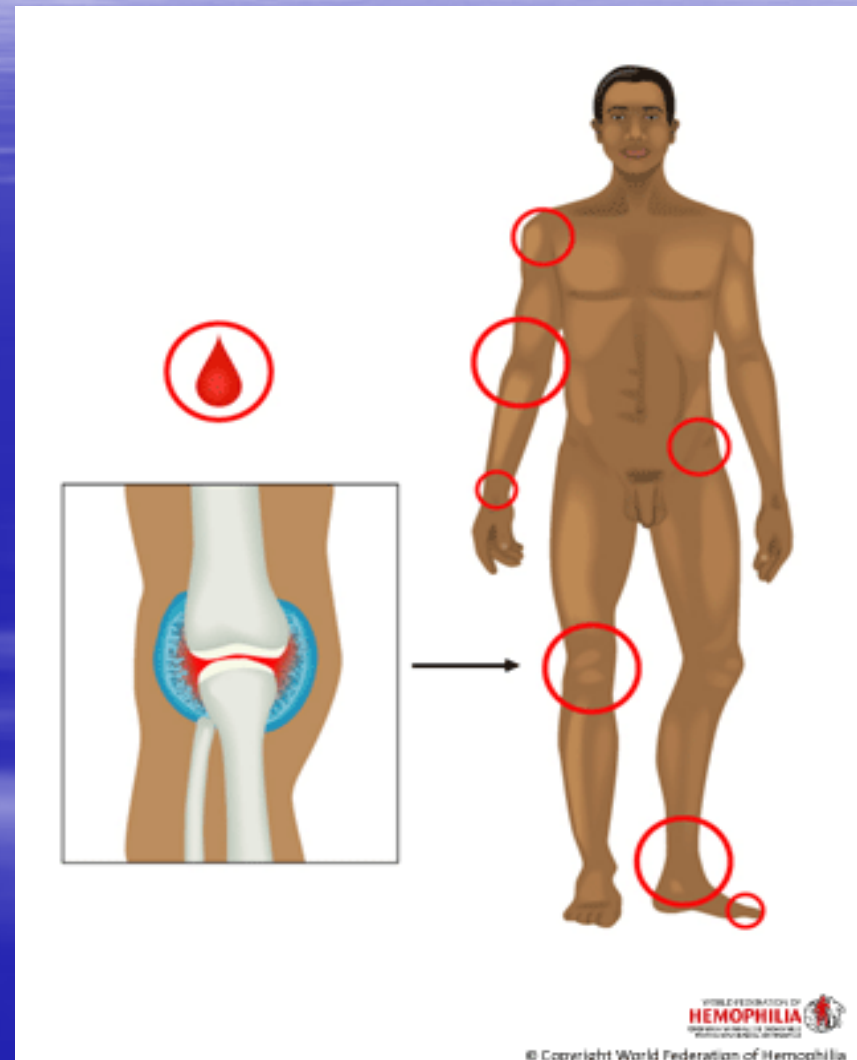
What happens in a joint bleed?

- A person with haemophilia knows when a bleed starts because the joint feels **tingly** and **warm**.
- As blood fills the capsule, the joint **swells** and becomes **painful** and **difficult to move**.
- Without treatment, the pressure from the swelling eventually stops the bleed. Later, special cells clear most of the blood out of the joint.



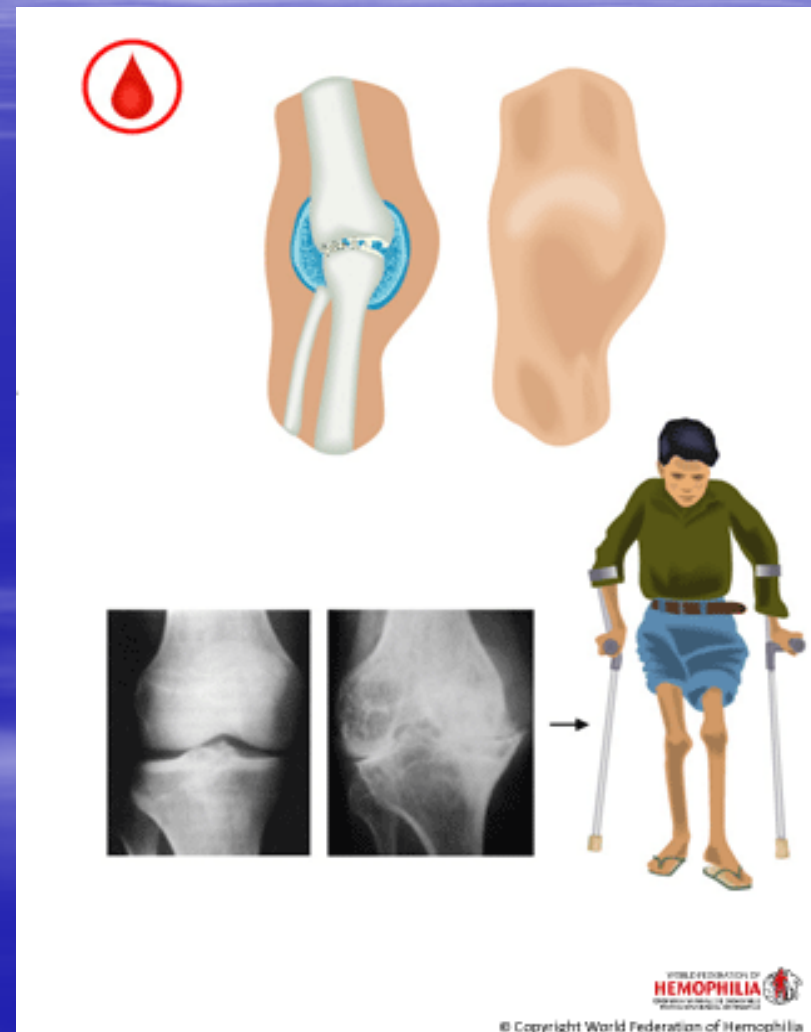
Which joint bleeds are most common?

- The most common joint bleeds happen in ankles, knees, and elbows – weight bearing joints
- Bleeds in other joints can also happen, including the toes, shoulders, and hips.
- Joints of the hands are not usually affected except after injury.



Long-term effects of Joint bleeds?

- Repeated bleeding into a joint causes synovium (lining) to swell and bleed very easily.
- Some blood remains in the joint after each bleed. The synovium stops producing the slippery, oily fluid that helps the joint move.
- This damages the smooth cartilage that covers the ends of the bones. The joint becomes stiff, painful to move, and unstable. It becomes more unstable as muscles around the joint weaken.
- With time, most of the cartilage breaks down and some bone wears away. Sometimes the joint cannot move at all. The whole process is called **haemophilic arthritis**.



Potential Outcomes

- Repeated bleeding / Inadequate or late treatment ⇒ more pain ⇒ immobility ⇒ muscle wasting and weakened joint ⇒ more susceptible to bleeding
- Vicious cycle of bleeding and re-bleeding - “TARGET JOINT”
- **CHRONIC ARTHRITIS** – a result of repeated bleeding and inadequate or late factor replacement.
- **JOINT REPLACEMENT!**

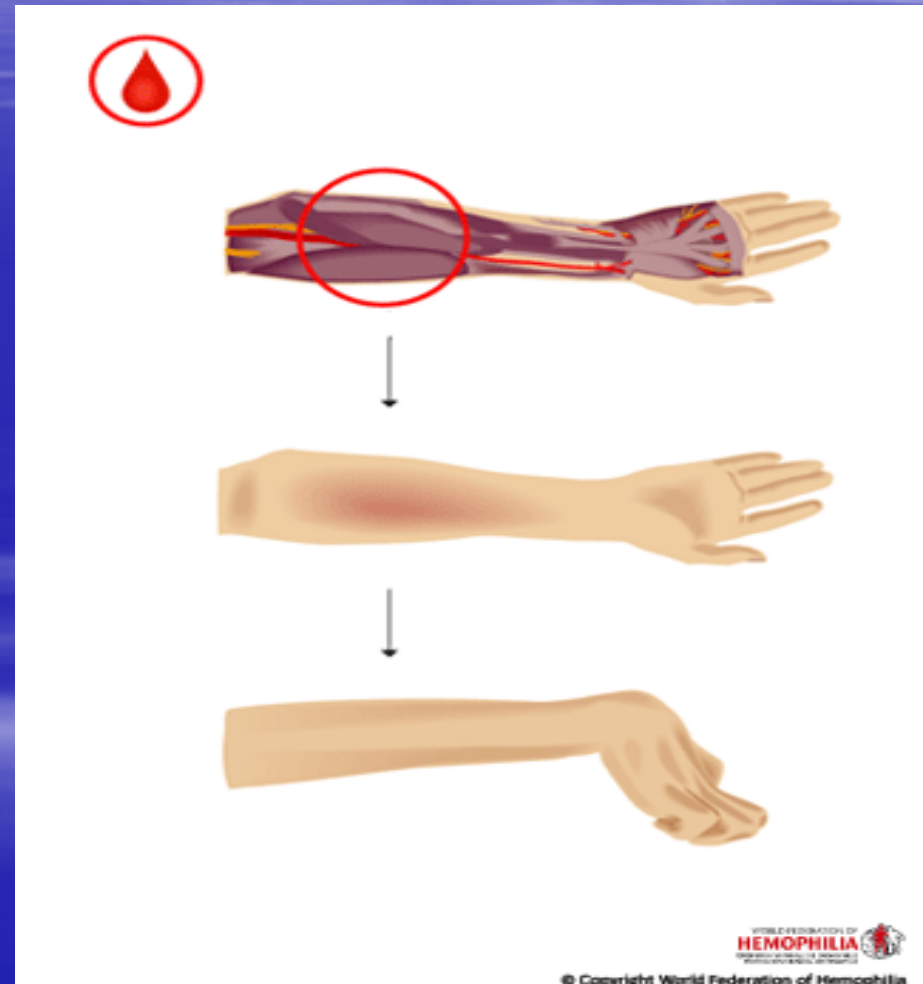
What Causes a Muscle bleed?

- Muscle bleeds happen when capillaries in the muscle are injured.
- Sometimes the cause is known, but bleeds can also happen for no clear reason.



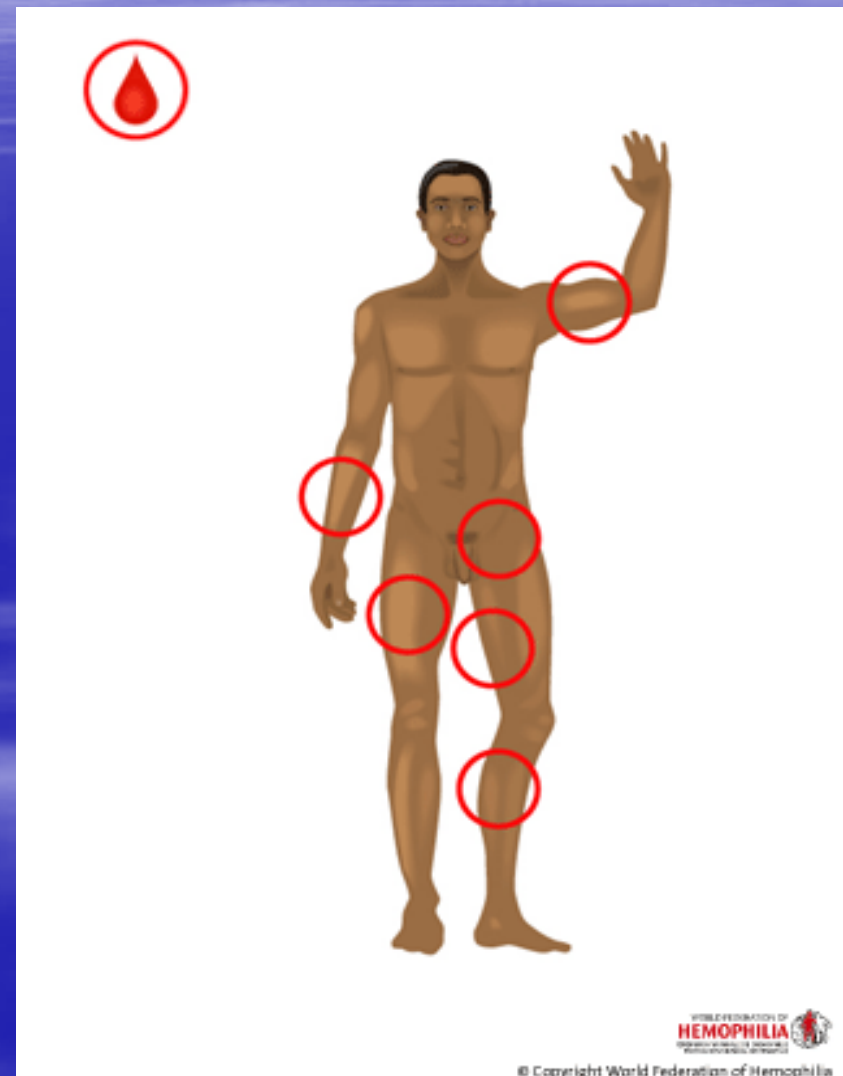
What Happens in a Muscle bleed?

- During a bleed, the muscle feels **STIFF** and **PAINFUL**.
- The bleed causes **SWELLING** that is **WARM** and **PAINFUL** to touch. There may be bruising if the bleed is near the skin.
- In some deeper muscles, the swelling may **press on nerves or arteries**, causing **TINGLING** and **NUMBNESS**
- The muscle **TIGHTENS UP** to protect itself. This is called a **MUSCLE SPASM**. As a result, joints usually moved by that muscle can't move properly.



Common Muscle Bleeds

- Calf, thigh, and upper arm.
- Bleeds in the **psoas muscle** (front of the hip) and the **forearm muscles** - also common.
Can put pressure on nerves and arteries, causing permanent damage. (numbness – classic sign)
- Bleeds into muscles of the hands are rare - usually follow injury.



Long-term effects of Muscle bleeds?

- After repeated bleeds, muscles can become **weak, scarred, and shorter** (sometimes permanently). **Can no longer protect joints.**
- Joints above and below the muscle can't move properly. **May bleed more often.**
- If nerves are damaged during muscle bleeds, the muscle may become weak or even paralysed.
- Permanent damage to joints, muscles, and nerves affects **sitting, standing, and walking.**



Spontaneous Bleeds

- Bleed without history of injury
- Target joint
- Importance of prophylaxis
- Importance of physiotherapy

Treatment

- Once a bleed has started it is important to raise factor level promptly to stop the haemorrhage
- Replacement should be continued until bleeding has stopped
- Further treatment - may be necessary to prevent re-bleeding during mobilisation and physiotherapy

Factor Concentrates

CONCENTRATE	INDICATION	HALF - LIFE
Advate Recombinant	FVIII Deficiency	12 Hours
BeneFIX Recombinant	FIX Deficiency	18 Hours
Novoseven Recombinant	Patients with inhibitors to FVIII or FIX	+/-2.7 Hours

Role of factor concentrate

- Replaces missing factor
- Injected IV
- Bleeding stops when enough factor reaches the bleeding site
- Treat ASAP

Treatment

- **A BLEED or POTENTIAL BLEED REQUIRES IMMEDIATE TREATMENT**
- If in doubt manage as a bleed at home and contact haemophilia team.

Formula For a Factor Rise

FVIII

- ✓ Recombinant Factor VIII increases the level two-fold (K=2)
- ✓ Dose Required = $\frac{\text{rise required (\%)} \times \text{weight (kg)}}{K (2)}$

Example:

Wt. 78kg, Rise 100%, Baseline level <1%

$$\frac{(100 - 0) \times 78}{2} = \frac{100 \times 78}{2} = \frac{7,800}{2}$$

= 3,900iu round up to 4,000iu

Formula For a Factor Rise FIX

- ✓ Recombinant FIX increases the level by 0.8 (K=0.8)
- ✓ Dose Required = $\frac{\text{rise required (\%)} \times \text{weight (kg)}}{K (0.8)}$

Example:

Wt. 65kg, Rise 70%, Baseline level <1%

$$\frac{(70 - 0) \times 65}{0.8} = \frac{4550}{0.8} =$$

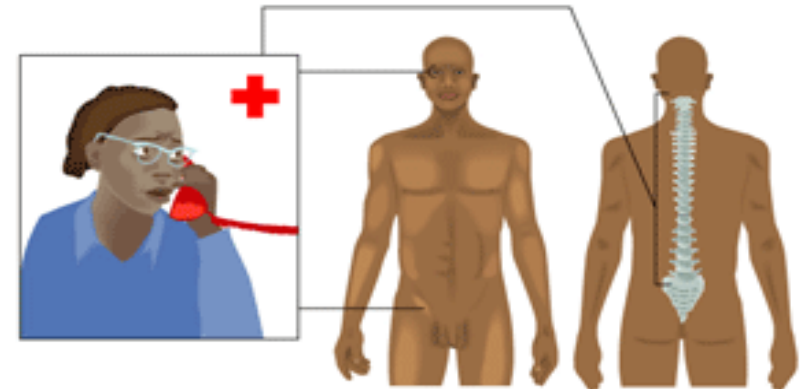
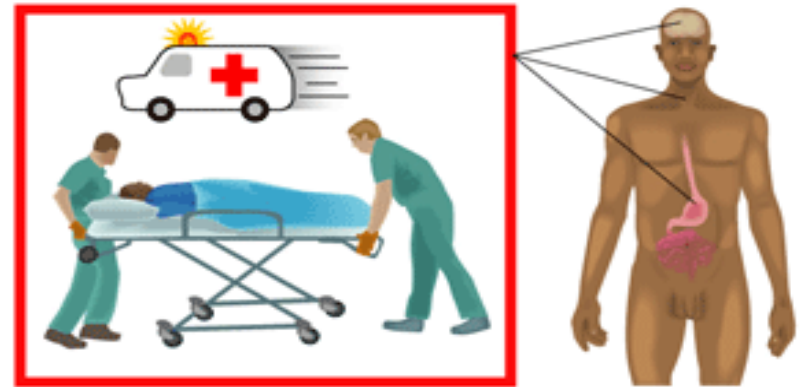
5687.5 rounded down to 5,500iu

Treatment Sheet

- Full name / Date of birth
- Date (including year), time (24hour clock)
- Reason for treatment (Bleed (site) / Prophylaxis)
- Total number of units and number of vials (Batch)
- Document Product administered and any comments

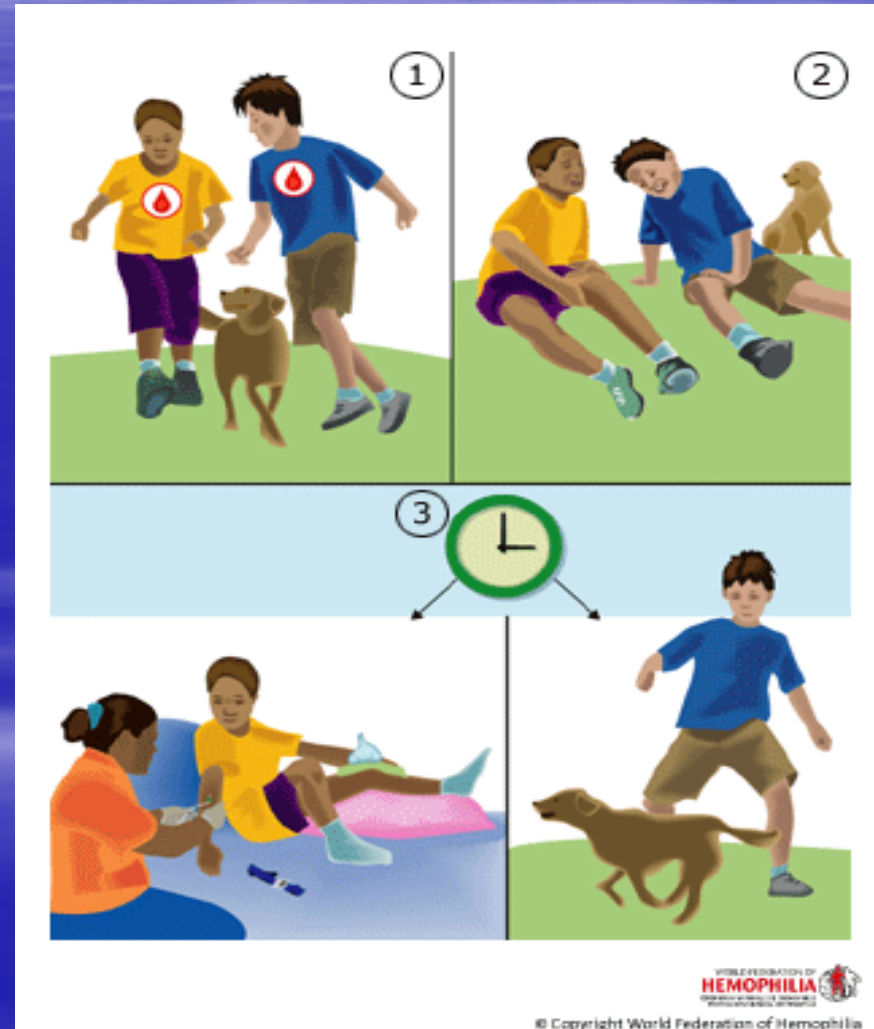
Serious or Life-threatening bleeds?

- Head injury - a major cause of death in haemophilia, esp. in children. Signs: headache, nausea, vomiting, sleepiness, confusion, clumsiness, weakness, fits, and LOC.
- Throat bleeds may result from infection, injury, dental injections, or surgery. Swelling, difficulty swallowing and breathing.
- Major blood loss uncommon except after injury or if related to another medical condition.
- Other bleeds may be very serious, but usually not life-threatening, eg bleeds into the eyes, spine, and psoas muscle.
- Blood in urine - common in severe haemophilia, but rarely dangerous.



Treatment of Bleeds

- Bleeds should be treated quickly to prevent later damage.
- If in doubt, treat. Don't wait!



Why treat quickly?

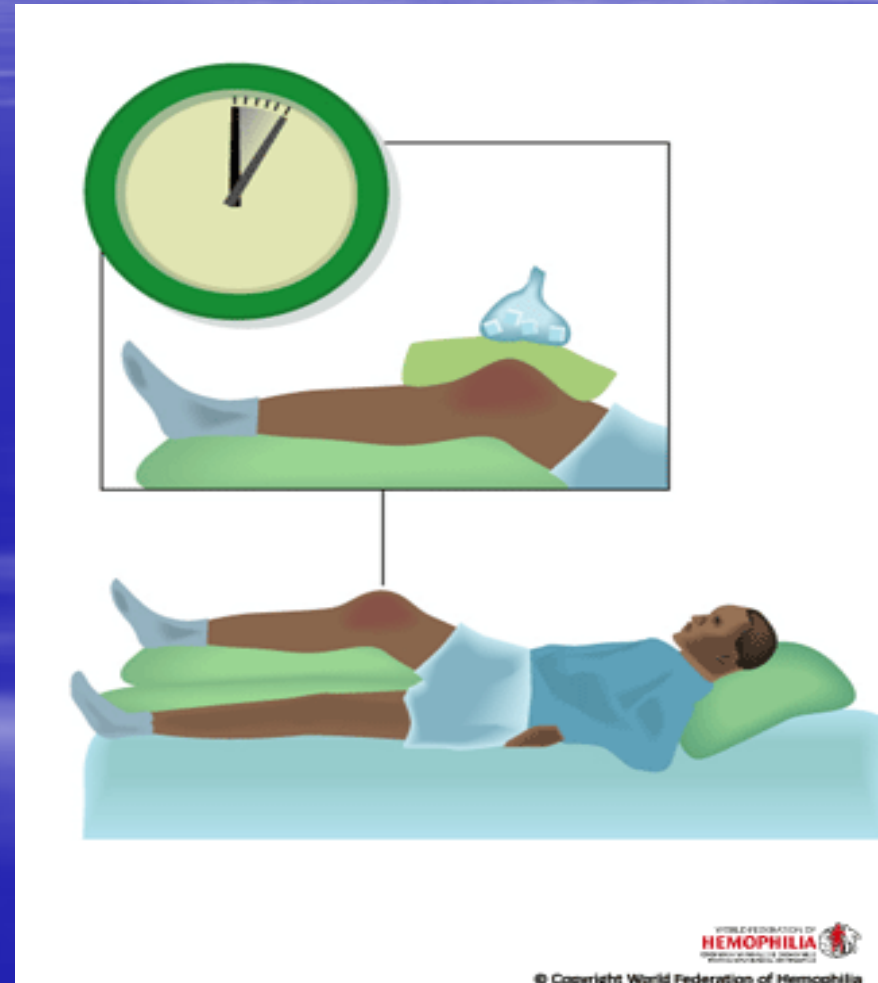
- When treatment is late, the bleed takes longer to heal, and treatment is needed for longer.
- More likely to have unwanted long term effects.



FIRST AID

R.I.C.E

- **REST:**
- **ICE:**



Other treatments that may help

- - Pain medication
- Review of factor replacement.

- **A physiotherapist can:**
 - Suggest ways to strengthen muscles and restore joint movement;
 - Say when it is safe to return to normal activities; and
 - Suggest ways to prevent further injury



Cuts and Abrasions

- Small cuts – do not cause a problem. Cover with plaster and / or bandage and apply pressure for a few minutes.
- Deep cuts – May need suturing and factor replacement to be given by family member / GP / A&E.
- Nose bleeds - pressing on nostril for 10 – 20 minutes or ice- pack on bridge of nose up to 5 minutes. - *Keep child sitting up and Do not put the head back.*
- If unsuccessful contact parent or haemophilia nurse specialist.

AWARENESS

- Prophylaxis
- Be aware, the younger child may be unfamiliar with what a bleed feels like. They may be upset, protect their limb by limping or avoid using it.
- Older children will inform you.
- Some need to miss school when recovering from a bleed, and may need some help in catching up. Or may come to school on crutches.
- Trusting relationships are hugely important.

AWARENESS cont'd

- All joint bleeds need to be treated immediately/ apply ice pack / arrange for administration of Factor / transfer to hospital if factor not available.
- Contact family or GP
- Superficial bruising is common. If it does not threaten function or mobility it does not need to be treated.
- Should be allowed to play alongside other children.
- Allow scissors etc., they need to learn how to handle them just the same as other children.

Inhibitors- What are they?

- **Inhibitors** - antibodies (proteins) made by the body to fight off things it sees as “foreign” – The body ‘neutralises’ infused factor
- Incidence ↑ with family history
- Majority <10yrs
- ↑ Bleeding

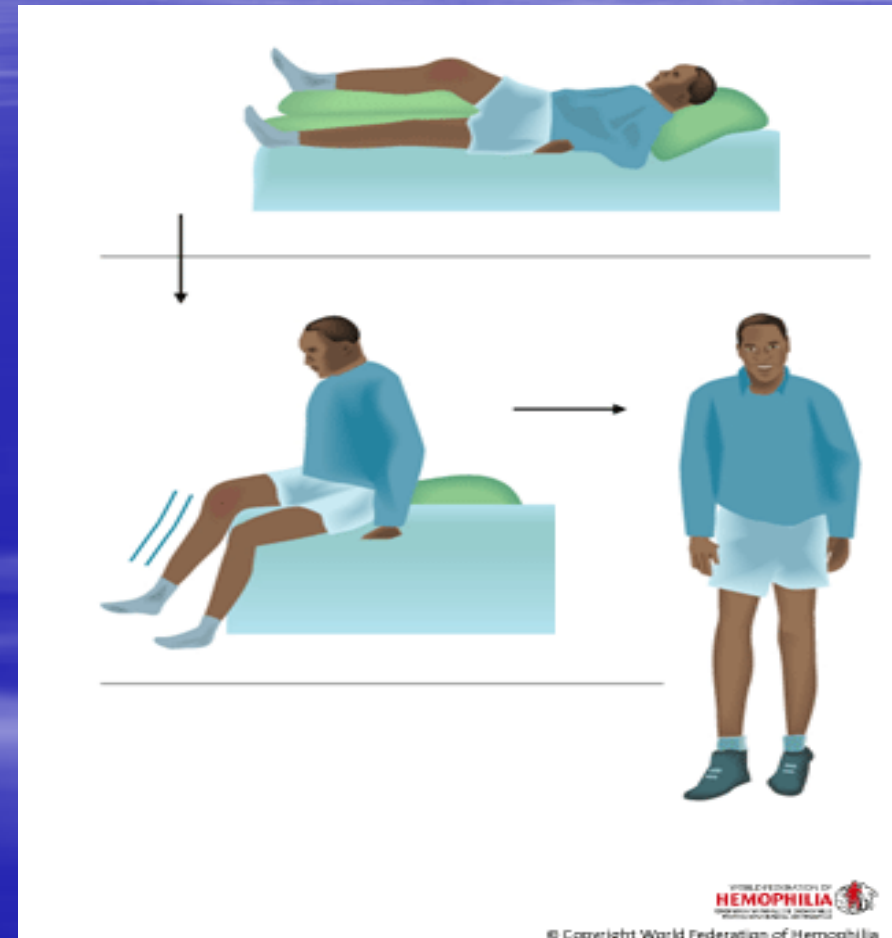


Signs of Recovery from a bleed

- Full movement of the joint or muscle returns

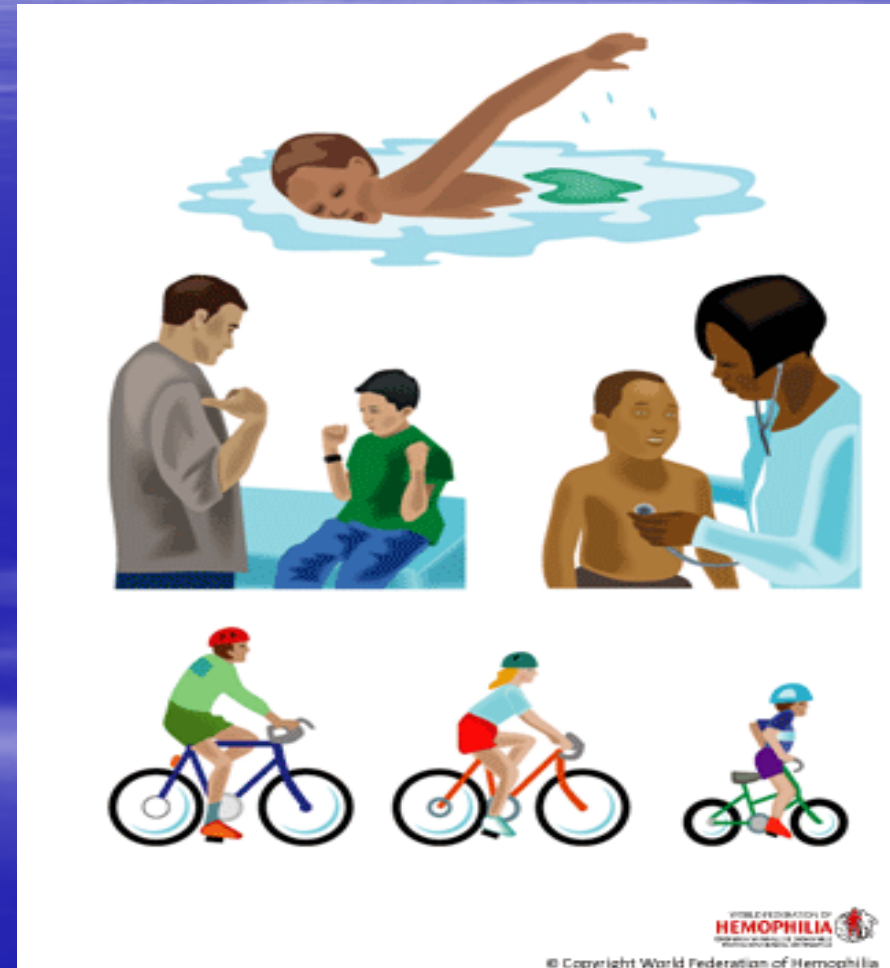
and

- Full strength of the muscle returns



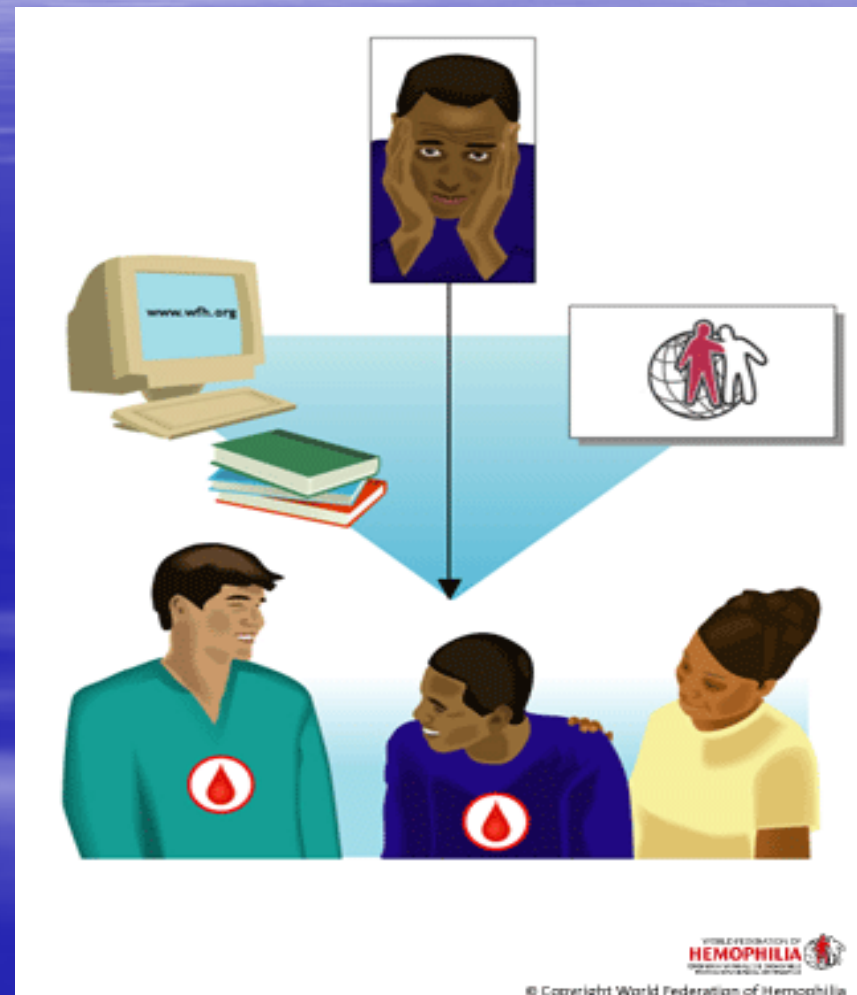
STAYING HEALTHY

- Exercise and stay fit
- Wear protection gear
- Regular check-ups, including joint and muscle examination
- Maintain a healthy body weight
- Vaccinations, hepatitis A and B protection



Staying Healthy

- Why is emotional health important?
- Living with haemophilia causes stress
- Learn as much as possible about haemophilia.
- Meet other people with haemophilia.
- Join a haemophilia society



Sport

- **Important to develop strong joints and muscles.**
- **As a rule avoid rugby, hurling and boxing.**
- **In primary school, sport is less competitive, therefore fewer problems.**
- **Time will tell if an activity provokes bleeds.**
- **Replacement therapy before games**
- **Warm up and cool down.**

Why is dental health important?

- Healthy teeth and gums reduce the need for haemophilia treatment.
- Regular dental care reduces the need dental surgery.
- Dental care should include brushing, flossing, and check-ups by a dentist and dental hygienist.



What if an operation is needed?

- Inform medical team
- Test for inhibitors
- Treatment plan
- Cyclokapron /
Tranexamic acid



Allergic Reaction

- What are some allergic reactions to treatment?
- Some treatments can cause an allergic reaction that may result in:
 1. Fever
 2. Shivering
 3. Skin rash
- Reactions are usually mild. They can be eased by taking **antihistamines** (medicine that is usually taken as pills).
- Medical help is needed quickly for:
 1. Difficulty breathing
 2. A tight feeling in the chest



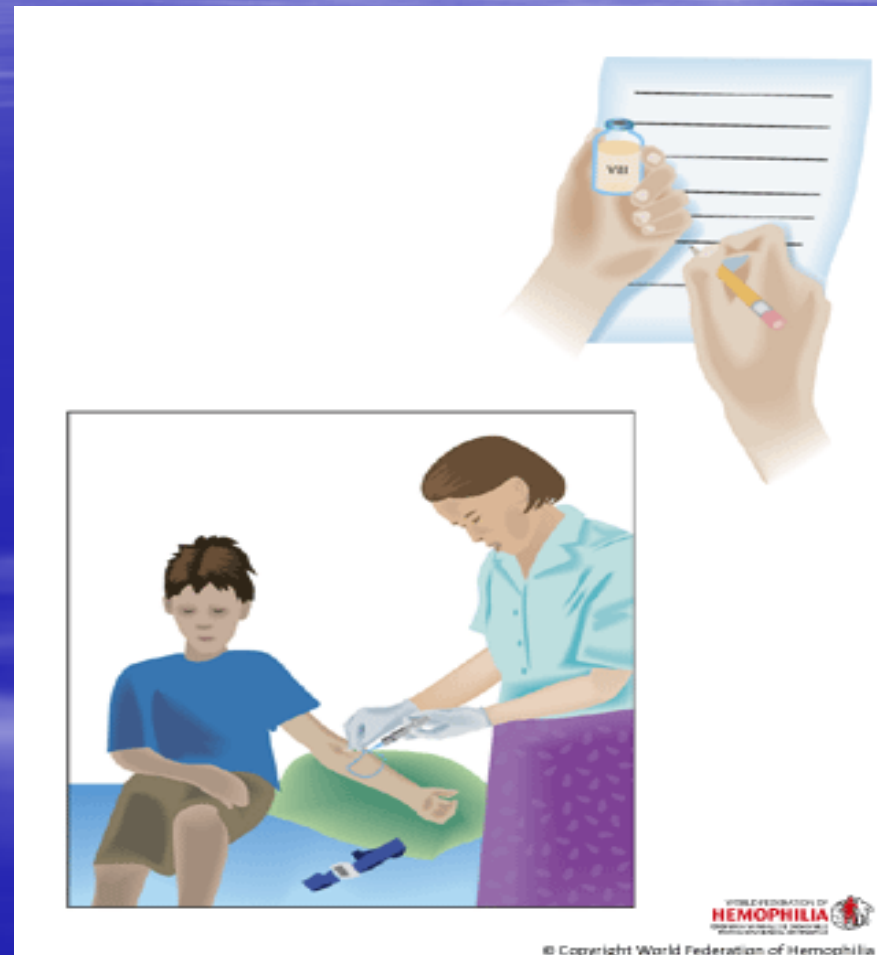
Identification

- A person with haemophilia should carry information about his health, including the type of haemophilia, treatment needed, and allergies (can be bought from chemist or on line)
- Business Cards



Home Therapy

- **Home therapy is infusion** with clotting factor replacement away from the hospital either IV / portacath.
- A written record of all treatments must be kept.
- Home therapy does not replace a doctor's or nurses care.



Venepuncture

- **Venepuncture** means to insert a needle into a vein.
- The best veins are usually seen or felt. Veins that are normally used are on the back of the hand or inside the elbow.
- Training is provided



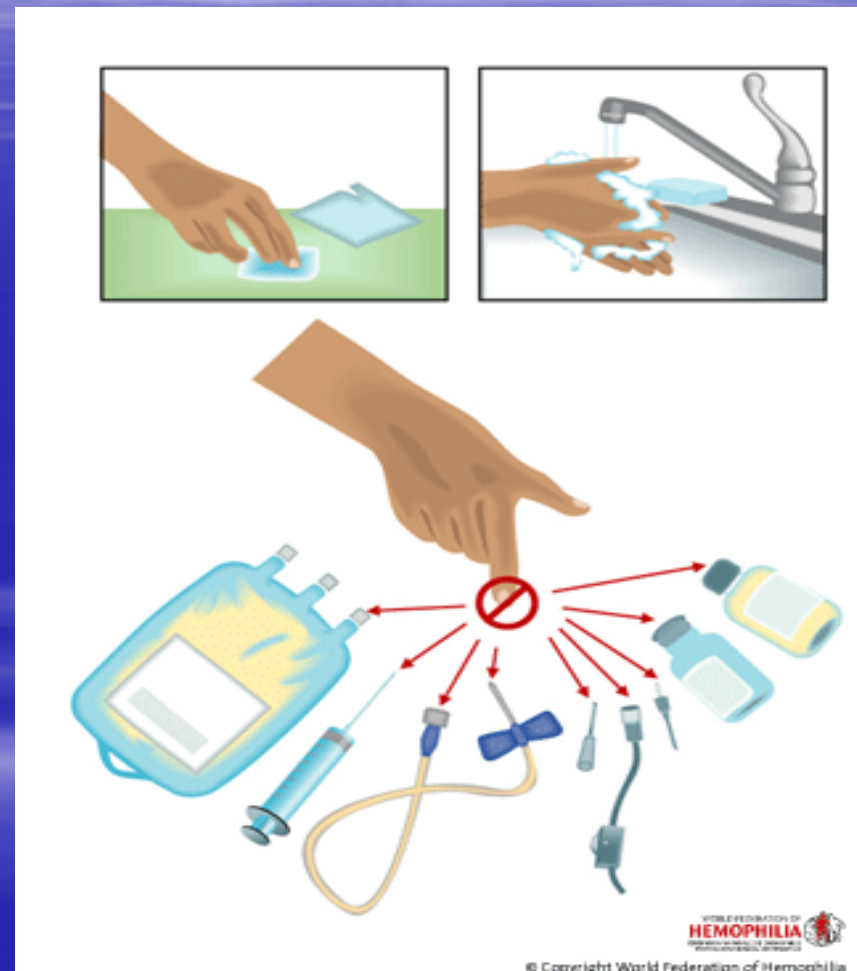
Preparation of Factor

- What preparation is needed for replacement therapy?
- Before handling any materials, wash hands thoroughly with soap and water. The work area should be cleaned with disinfectant.
- Insertion of needle into Port-a-cath should be under sterile procedure



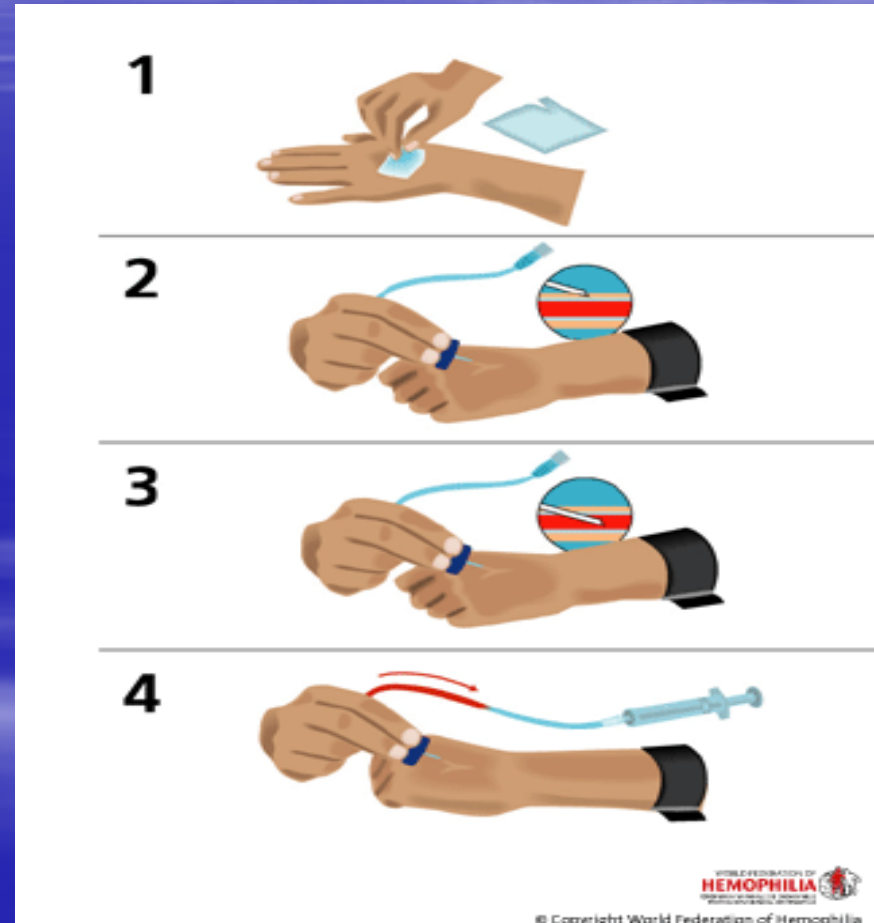
Prevent Infection

- How can infection be prevented when giving an injection?
- Aseptic technique
- Sharp objects
- Concentrates should not be opened and then stored for later use.
- Clean any spills with disinfectant.



Factor Replacement

- Be sure that your hands are clean.
- Use all of the dissolved concentrate.
- Put pressure on site after removal of needle for 5mins



What is a Port?

A Port is a generic term that describes any total implantable central venous access device (TICVAD) which is positioned within a vein and provides IV access

It consists of

- A fluid reservoir with an artificial self-sealing septum
- catheter (a thin, flexible hollow tube)

Port-a Cath's



Indications

- Long term regular administration of IV drugs, blood products or fluid therapy
- Multiple blood testing
- Poor IV access

Advantages of Port's

- Reliable IV access
- Avoids thrombophlebitis of peripheral veins associated with regular veno-puncture and infusions
- Increases comfort and reduces anxiety for patients
- Lower infection rate
- Physical activity is not restricted

Disadvantages of Port

- Surgical procedure
- Risk of complications
- Needle required to access the device
- Risk of needle dislodgment
- There will be scarring and a small bulge where the reservoir is located

Complications associated with the surgical procedure

- Bleeding
- Pneumothorax/Haemothorax
- Arterial puncture
- hematoma
- air embolism,
- cardiac arrhythmia

Complications associated with the Port

- Infection
- Catheter fracture
- Occlusion fibrin sheath formation/catheter thrombosis/kinking
- Venous thrombosis/pulmonary embolis
- Air embolis
- Endocarditis

Port



Position



Needle in Situ



Advice

Where can people get help or advice about haemophilia?

- Hemophilia healthcare workers
- Hemophilia organizations www.haemophilia-society.ie and www.wfh.org
- Other people with haemophilia and their families (through meetings etc)
- Literature



Finally

- Children with haemophilia can lead a normal life and should not be treated any differently to other children.
- When a bleed occurs, early treatment is crucial, which can prevent long-term complications.
- If in doubt, contact the child's family, their GP or the nearest haemophilia centre.
- Contacts details
- Go raibh mile maith agat agus slán

References

- WFH – World Federation of Haemophilia – Haemophilia in Pictures www.wfh.org
- Haemophilia Foundation of Australia
“Finding out about the haemophilia gene”
- Mary Kavanagh OLCHE “What is a Port”

Contact Details

- Hereditary Bleeding Disorders Nurse Specialists

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Mobile: 087-9683246 (Emergency only)

After 5pm and weekends:

Haematologist on call 021-4546400

