BASICS OF HAEMOPHILIA

Introduction

Haemophilia is a genetic bleeding disorder in which there is a deficiency of clotting factors in the blood. Due to the deficiency of these factors, blood is unable to clot itself and the disease is known as haemophilia.
Approximately 10000 to 15000 people in Pakistan are diagnosed with haemophilia. 70% of these patients acquire haemophilia genetically whereas 30% of the patients have no family history of haemophilia. It is usually a male dominated disease and is transferred from unaffected women who are its carriers.
There are two types of haemophilia: Haemophilia A and Haemophilia B. Haemophilia A is factor VIII deficiency and Haemophilia B is factor IX deficiency.
Patients with haemophilia have problems clotting their blood even after minor scrapes or injuries. It can take weeks to clot blood for some patients and for some patients the blood clots within a few hours. If pressure is applied on the affected area immediately, it clots blood right away.

Spread of haemophilia and its affect on blood

Haemophilia is a genetic disease and does not spread like a virus. It is transferred to the child by parents. In some cases genetic changes in the body can also cause haemophilia. But almost 70% of the patients acquire this disease through parents. This is called sporadic haemophilia.
Factors help to clot blood and are present in the human genes. If there is some problem in these genes, then future generations will also have the same genetic problem.
Haemophilia A and B are both related to X-Chromosomes. This is a male dominated disease because they have only one X-Chromosome. Whereas women have two X-Chromosomes and therefore if there is a problem with one, the other can cover it up.

Symptoms

- Bruising on the body
- Spontaneous bleeding
- Prolonged bleeding after surgery, injury or dental treatment
- Continuous bleeding after an accident or head injury
- Swelling and imbalance in the joints
- Warming of the affected joint and muscle

Different types of haemophilia

1. Mild Haemophilia (5% - 50% clotting factors)

In mild haemophilia, the level of factor VIII is between 5% and 50%. Following are its symptoms:

- Prolonged bleeding after surgery
- No spontaneous bleeding

2. Moderate haemophilia (1% - 5% clotting factors)

In moderate haemophilia, the level of factor VIII and IX is between 1% and 5%. People with moderate haemophilia can bleed after minor cuts or injuries. For example:

- Heavy bleeding during surgery, injury or dental treatment
- Bleeding episode once a month
- Spontaneous bleeding

3. Severe Haemophilia (factor level < 1%)

In severe haemophilia, the level of factor VIII and IX is less than 1%. Its symptoms are:

- Internal bleeding in joints and muscles
- Bleeding episode once or twice a month
- Spontaneous bleeding

Diagnosis

If you have any of the above symptoms, contact your nearest haemophilia center. If the doctor feels you have haemophilia, he will advice you some of the following tests:

- PT
- APTT
- Factor level

Blood tests are important because you can measure the clotting time through them. Symptoms of haemophilia can be different in patients. People with mild or moderate haemophilia also complain about bleeding. Women who are carriers of haemophilia can find out in first 10 – 12 weeks of their pregnancy through DNA testing that whether their child has haemophilia or not.

Treatment

The treatment of haemophilia depends on the factors deficient in the blood. For example, if a patient has factor VIII deficiency, he will be injected with a required amount of factor VIII which will stop the bleeding. In the same way in factor IX deficiency, factor IX is injected. If factors are unavailable, FFP and cryoprecipitate can also be given. Cryoprecipitate is no good in factor haemophilia B.

When are factors needed?

- Swelling in joints
- Injury to mouth, nose, tongue etc
- After heavy and prolonged bleeding anywhere in the body
- After an accident or a surgery

In what situations factors/plasma should be given before treatment?

- Operation or dental treatment
- Any activity which involves the risk of bleeding (for example: circumcision, surgery)

Preparation of factors

- Remove the cover from the small needle
- Insert the needle into the bottle with water
- Remove the cap from the factor bottle and put water in it
- Now shake it gently to mix water and factor otherwise factor will be spoiled in foam
- Put the filter and take 10cc factor in the syringe
- Use butterfly needle to inject the factor in the veins
- Be careful not to waste any factor
- Remove the needle carefully and put a plaster

Safety precautions

Do's

- Put cool wraps on the affected joint or muscle in the first 24 – 48 hours in case of swelling or pain
- Prefer indoor games such as ludo, video games, scrabble etc
- Take good dental care and always use a soft toothbrush
- Use paracetamol in pain
- Always use a lift to go upstairs
- Keep regular contact with the school staff of your child so they are aware of your child's condition
- Girls should use haematonics regularly

- Regular checkups
- Contact your haemophilia center in heavy menstrual periods
- Use transamine capsule one day before menstruation

Don’ts

- Avoid using hot wraps in pain and swelling
- Do not use aspirin
- Sports like football are dangerous and should not be played
- Avoid intramuscular injections.
- Don’t use hard toothbrush
- Avoid climbing stairs
- Keep sharp objects away from children
- Never leave the child alone
- Avoid physical exertion

Note: consult your doctor before using any medicine

Home care

Ice treatment

- Put ice in a clean cloth and crush it
- Put ice on the affected area for 5 minutes
- Give 10 minutes interval and repeat the process

Note: ice treatment is very beneficial in severe pain and swelling but it should be done in first 48 hours.

How to use analgesics

Analgesics should be used in case of pain in muscles or joints and in schools and offices. Analgesics should be used according to the table below:

Name of drug	Dose for adults	Dose for children
Paracetamol	1 – 2 tablets every 4 – 6 hours	1 tea spoon of syrup
Tramadol	1 – 2 capsules every 6 hours	Not recommended for children

Rehabilitations

Patients with haemophilia should know the importance of physiotherapy once the swelling or pain has decreased. Physiotherapy strengthens muscles and bones and reduces the probability of future bleeding.

Dangerous bleeding areas

Bleeding into head, mouth, throat and intestines is extremely dangerous and the patient should immediately contact the nearest haemophilia center.

Use of transamine in dental bleeds

- Mix one tablet (500 mg) in one tablespoon of water
- Gargle with the solution or try to keep it in mouth as long as possible
- Swallow the solution
- For kids, mix it in little water to form a paste. Then apply the paste on the affected area

In case of nose bleeds

In case of nose bleeds, use a bandage soaked in injection transamine and put it inside your nose. Once the bleeding has stopped, dampen the bandage and pull it out carefully. For kids who have regular nose bleeds, apply Vaseline or petroleum jelly in the nose regularly.