HEMOPHILIA AND ROLE OF A NURSE

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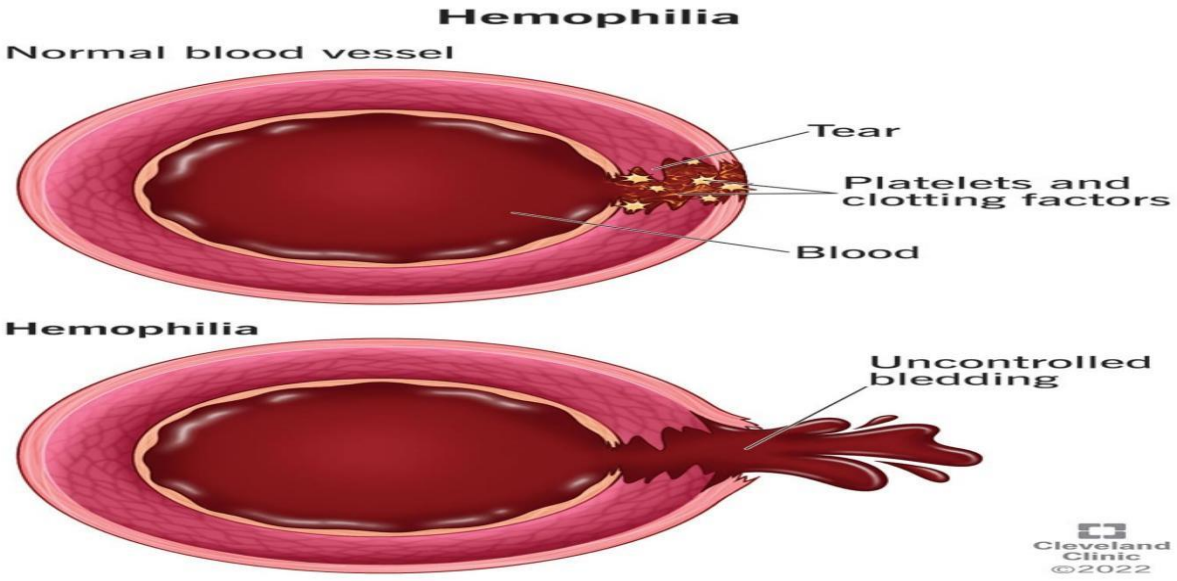
Abstract

Haemophilia is a genetic bleeding disorder where blood doesn't clot due to lack of enough blood-clotting proteins. In case of injury haemophiliac patient bleed for a longer time than a normal person, even small can cause problem among them. Haemophilia is mainly concerned with the bleeding I.e internal bleeding, especially knees, ankles and elbows that damages organs, tissues and can be fatal.

Key words: Haemophilia, bleeding, organs ,genetic disorder, clotting factor.

Introduction

Hemophilia is a genetic bleeding disorder where blood doesn't clot in a way it should be due to lack of blood-clotting proteins .People with hemophilia have low levels of either factor Factor VII(7), VIII (8) and factor IX (9).This condition is associated with automatic bleeding as well as bleeding after injuries or surgery. The severity of Hemophilia (severe, moderate or mild) is based on the amount of clotting factor present in a blood.Hemophilia typically affects men rather females ,rarely certain females may have low clotting factors and can develop symptoms of very heavy periods.



Types of hemophilia:

Hemophilia based on type	Level of hemophilia based on severity
Hemophilia A develops when there is low level of factor	Mild hemophilia people having 5% to 30% of the

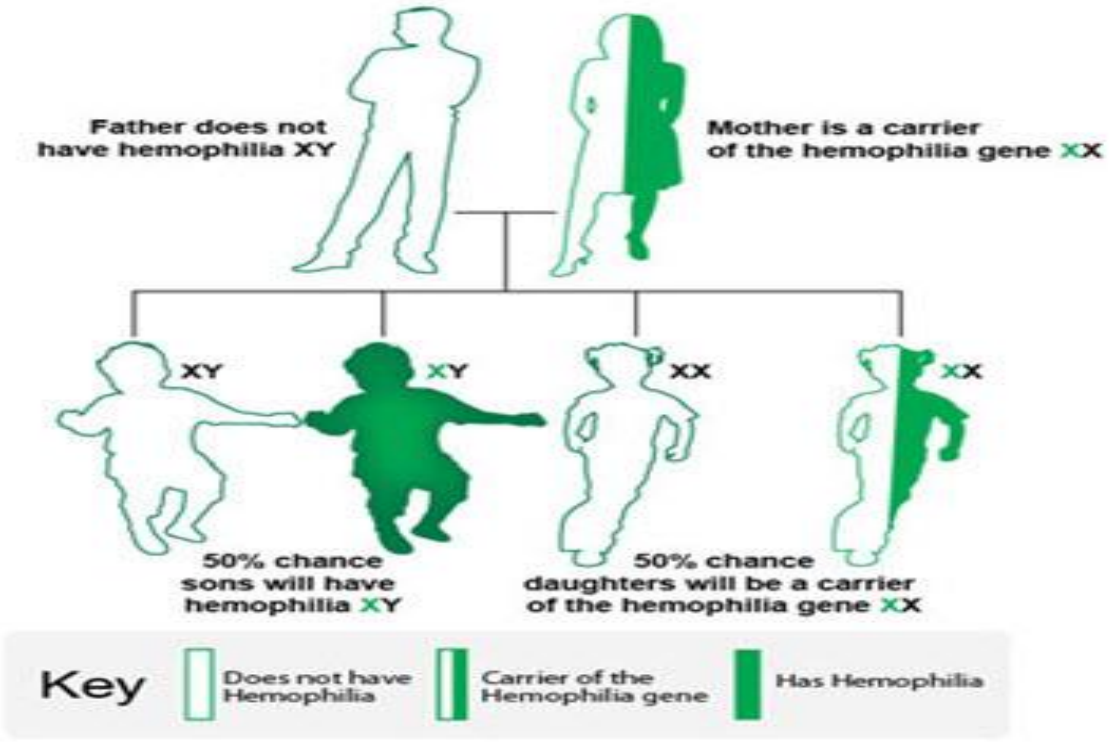
VIII .It is thecommonest type of hemophilia	normal amount of clotting factors in their blood .
Hemophilia B develops when a person don't have enough clotting factor IX.	Moderate hemophilia people with 1% to 5% of the normal level of clotting factors.
Hemophilia C is also known as factor XI deficiency. This is very rare	Severe hemophilia people with less than 1% of the normal clotting factors .

Symptoms and Signs

General Symptoms and Signs	Symptoms and Signs Babies and Children
Symptoms of Hemophilia are significantly unusual or excessive bleeding or bruises . Level of bleeding or bruises depends on whether patient is having severe,moderate or mild haemophilia	Babies or Children may bleed more than usual after circumcision and common symptoms among children are :
Patients developbruises soon after minor injuries& is a sign of bleeding under the skin. Unusual bleeding for a longer time may be after surgery ,simple cuts or dental treatment. Bleeding without any reason like sudden nose bleeding. Internal Bleeding within the joints (ankles, knees, hips and shoulders)& Brain causing Joint pain, persistent headache, double vision, swelling feels hot to touch as well	Bleeding and swollen lumps among babies or toddlersmay occur from mouth after minor injuries like bumping during their oral gratification. They also refuse to crawl after muscle or joint internal bleeding.Their bodies look bruised and swollen and feel warm to touch Hematoma is a mass of congealed blood that is accumulated under the skin usually after receiving an injection.

Etiology/Pathophysiology

Hemophilia is inherited disorder caused by mutation or change in genes that give instructions to clotting factor proteins to form a blood clot. If a woman has an abnormal factor gene on one of their X chromosomes, they carry hemophilia but may not have symptoms. That's because there's a normal factor gene on their second X chromosome.If a woman who carries an X chromosome with a defective gene for producing factor 8 (or factor 9) has a male child, that child has a 50% chance of inheriting the X chromosome that carries the abnormal factor gene..Thus it is X linked disease that is how males are mostly affected and females are mostly carriers .Even though hemophilia runs in families, some families have no prior history of family members with hemophilia. Sometimes, there are carrier females in the family, but no affected boys, just by chance.



Diagnosis

Hemophilia can be diagnosed by taking a complete personal and family history and physical examination. Certain blood tests are help in their Diagnosis

Complete blood count (CBC): to cheek and measure blood cells.

Prothrombin time (PT) test: studies how quickly blood clots.

Activated partial thromboplastin time test: This is another blood test to time blood clot formation.

Specific clotting factor test(s): This blood test show levels of specific clotting factor levels (such as factor 8 ,7and factor 9).

Medical Management

Hemophilia is managed by boosting clotting factor levels or replacing clotting factors.Blood transfusions are also needed. Replacement clotting factor can be made from donated blood. Similar products, called recombinant clotting factors, are made in a laboratory, not from human blood.

Other therapies include:

- ✓ Desmopressin. In some forms of mild hemophilia, this hormone can stimulate the body to release more clotting factor. It can be injected slowly into a vein or used as a nasal spray.
- ✓ Emicizumab (Hemlibra). This is a newer drug that doesn't include clotting factors. This drug can help prevent bleeding episodes in people with hemophilia A.
- ✓ Clot-preserving medications. Also known as anti-fibrinolytics, these medications help prevent clots from breaking down.

- ✓ Fibrin sealants. These can be applied directly to wound sites to promote clotting and healing. Fibrin sealants are especially useful for dental work.
- ✓ Physical therapy. It can ease signs and symptoms if internal bleeding has damaged your joints. Severe damage might require surgery.
- ✓ First aid for minor cuts. Using pressure and a bandage will generally take care of the bleeding. For small areas of bleeding beneath the skin, use an ice pack. Ice pops can be used to slow down minor bleeding in the mouth.

Nursing management/ Role of a nurse

- ✓ Give Medication that is factors as prescribed.

Health Education

- ✓ Educate the patients & their family that Bumps, falls and hard knocks may proven a serious issue ,so educate them to avoid activities that increase their risk of being hit or falling down after being hit like Playing football, hockey or rugby, boxing or wrestling, Riding motorcycles,Riding skateboards.
- ✓ Patients are advised not to take medications used for pain (ibuprofen) and anticoagulants such as heparin or warfarin.
- ✓ Educate the patient and family for Activities and actions that can improve quality of life
- ✓ Develop an exercise routine as advised.
- ✓ Manage their stress: Hemophilia is a lifelong illness. It may take extra effort to balance your obligations to your family and work.if stress hampers your quality of life ,take treatment from mental health professional.
- ✓ Have good dental hygiene by proper Brushing, flossing and regular visits to your dentist concerned.
- ✓ Weight management may reduce risk of other orthopedic problems
- ✓ Self infusion training should be done in order to make the patient self sufficient as recommended by World federation of Hemophilia.
- ✓ Provide supportive therapies .Infact refer them in case mental health issues they have developed or any other physical issues to the concerned.

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