Tikrit University

College of Nursing

Clinical Nursing Sciences

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Child Health Nursing

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(Bleeding Disorders (Hemophilia))

by:

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Bleeding Disorders (Hemophilia)

There is a deficiency of one of the factors (proteins) necessary for coagulation of the blood.

80% of all cases of hemophilia is X-linked recessive inheritance.

Certain blood cells and substances normally form clots and stop the child from bleeding too much. These include:

A. platelets B. clotting factors C. vitamin K D. fibrinogen.

Types of Hemophilia

1.**Hemophilia A or Classic hemophilia**: is commonest 80% of cases.

2.**Hemophilia B or Christmas disease**: deficiency of factor IX

Grades of Hemophilia

Hemophilia can be mild, moderate, or severe.

Depending on the amount of clotting factor or the kind of bleeding episodes the child has.

❖ Severe Hemophilia:

1) Factor 8 or factor 9 less than 1%
2) Frequent sudden spontaneous bleeding
3) (usually in the muscles or joints (Hemarthroses).
4) Bleeding with minor trauma.
5) Manifests in Infancy

❖ Moderate Hemophilia.

1) Factor 8 or factor 9 usually 1-5%
2) Require moderate trauma to induce bleeding episodes

❖ **In Mild hemophilia**

1) Factor 8 or factor 9 usually (>5%),
2) No spontaneous bleeding
3) may go undiagnosed until he has a tooth pulled, surgery, or a bad injury.
4) only have severe bleeding after a major surgery or severe injury.

Von Willebrand Disease (Vwd)

is another hereditary bleeding disorder characterized by a deficiency, abnormality, or absence of the protein called von Willebrand factor (vWF) and a deficiency of factor VIII.

vWD is an autosomal recessive, Unlike hemophilia, vWD affects both males and females.

1) factor VIII (antihemophilic factor [AHF]
2) AHF is produced by the liver and is necessary for the formation of thromboplastin in phase I of blood coagulation.
3) Bleeding into subcutaneous and IM tissue is common.
4) Hemarthrosis, which is bleeding into a joint space, is the most frequent type of internal bleeding.
5) Bony changes and crippling deformities occur after repeated bleeding episodes over several years.
6) Signs of hemarthrosis are swelling, warmth, redness, pain, and loss of movement.

7) Bleeding in the neck, mouth, or thorax is serious because the airway can become obstructed.
8) Intracranial hemorrhage can have fatal consequences and is one of the major causes of death.
9) Hemorrhage anywhere along the GI tract can lead to anemia, and bleeding into the retroperitoneal cavity is especially hazardous because of the large space for blood to accumulate.
10) Hematomas in the spinal cord can cause paralysis.

Clinical manifestations (S &S)

● Frequent bruising.

● Frequent bleeding from the nose or gums.

● Pain and swelling in joints or muscles that lead to limited motion.

● Bleeding that lasts a long time.

● Bowel movements that are black (melena).

● Urine that is pink or red (hematuria).

- Hemorrhage from any trauma—Loss of deciduous teeth, circumcision.

● Subcutaneous and intramuscular hemorrhages.

Diagnostic

1) Clinical history is crucial in the diagnosis
2) Family "tree" should be constructed in any case where there is a history of bleeding in other family members, and those particular aspects to look out for are: bleeding or lack of it after dental extraction, circumcision, tonsillectomy and child birth trauma.
3) Platelets count, bleeding time measurement.
4) Prolonged activated partial thromboplastin time (aPTT).

5) X ray for hemarthrosis.

Therapeutic management /Treatments of hemophilia

1. **Factor Replacement therapy**: Bleeding associated with surgery, trauma, or dental extraction often can be anticipated, and excessive bleeding can be prevented with appropriate replacement therapy.

2-Demand therapy: child receives clotting factor to stop bleeding episode.

3-Prophylactic therapy: To prevent future joint bleeding episodes.

For life-threatening bleeding, levels of 80-100% of factor 8 or factor 9 are necessary.

For mild to moderate bleeding episodes (hem arthroses), a 40% level for factor 8 & 9 is appropriate.

4. **Surgery**: Surgery may be done to repair any damage caused by bleeding in the child's joints.

5. **Gene therapy**

Drugs

1) **Anti fibrinolytic proteins:**
- keep blood clots from breaking down.
- used to stop bleeding in the child's mouth, nose, or abdomen**.**

2) **Desmopressin:**
- used to treat hemophilia A.
- works by increasing amount of clotting factor VIII

3) **Non-steroidal anti-inflammatory drugs (NSAIDs),**
- ibuprofen, are effective in relieving pain caused by synovitis
- occasionally used with caution because they inhibit platelet function.

Nursing management

1. Nursing intervention to prevent bleeding

● Complete physical assessment of the child with hemophilia

● Continuously assess the level of clotting factor

● Playing should prevent bleeding by emphasizing on type of toys (smooth (soft) not sharp).

● Encourage to walk and swim.

● Advise parents about types of sports for hemophilia

● advised to wear a helmet, a seat belt, and a protective strap over the knee and elbow area to protect them from injury.

● Replacing hardwood, ceramic and marble floors with rugs and carpets, and these precautions only reduce the risk of bleeding injuries.

● Keep an eye on every move that allows the occurrence of injury.

● Maintain security of the child bed.

2. Intervention during bleeding episodes

● **Rest**: Have the child sit or lie quietly until bleeding episode ends.

● **Ice:** Ice helps decrease swelling and pain. Ice may also help prevent tissue damage. Use an ice pack, or put crushed ice in a plastic bag. Cover it with a towel and place it on the area for 15 to 20 minutes.

● **Compression:** Apply pressure to the bleeding site. A bleeding joint can be wrapped with tape or an elastic bandage.

● **Elevation**: Position the child so the bleeding area of his body is raised. Raise the area above the level of his heart if possible. Prop the area on pillows to keep it elevated comfortably.

● Provision of blood transfusion, to repair red blood cell count and enhance oxygen-carrying capacity to be adequate tissue perfusion

3. Physiotherapy

4. Emotional support

Management of hemarthrosis

1. Observation of clinical signs and symptoms of hemarthrosis such as **hematoma, local pain, swelling, limited motion**.
2. Elevate and Immobilization of the joint with splint or traction
3. Control the degree of exercise according to degree of discomfort
4. Application of cold sponge.
5. Physiotherapy with careful handling to prevent bleeding.
6. Replacement therapy to increase the level of factor 8, plasma can be given also or blood transfusion
7. Give analgesic to control pain according to Dr