HEMOPHILIA

DEFINITION

- Hemophilia is a group of bleeding disorders resulting from congenital deficiency of specific coagulation proteins
- Hemophilia is due to congenital deficiency of plasma coagulation factor VIII or factor IX. Inheritance is X-linked recessive. The female carries the hemophilic trait but only the male offspring suffers from the disease.

CLASSIFICATION

- Type A (80%)- Deficiency of factor VIII.
- Type B- Also known as Christmas disease. Caused by lack of factor IX.
- Type C-Caused by a lack of factor XI. It is an autosomal dominant or a complete recessive traits.
- Type D (Von Will brand's or vascular hemophilia): Caused by decreased levels of factor VIII (Factor VIIIag & VIII levels are both depressed). It is an autosomal dominant trait.

CLINICAL

Mild & moderate.H-

Asymptomatic . May develop prolonged bleeding following tooth extraction, severe trauma or following surgery.

Severe.H-

Excessive bleeding.

Subcutaneous and intramuscular bleeding

Hemarthrosis

Epistaxis

Retroperitoneal bleeding

Intracranial bleeding

DIAGNOSTIC EVALUATION

- Family history
- Bleeding time
- Clotting time
- Partial thromboplastin time
- Factor VIII or IX assay
- DNA test

MEDICAL MANAGEMENT

- Replacement of the missing clotting factor

  -One unit of plasma(250ml) given daily-Commercial preparation of factor VIII(1-deamino-8-D-arginine vasopressin
- Corticosteroids
- NSAID-Ibuprofen
- Epsolon-aminocarporic acid
- Fibrin glue

NURSING MANAGEMENT

- Prevent bleeding
- Recognize and control bleeding
- Prevent crippling effects of bleeding
- Support the family and prepare for home care
- Antenatal diagnosis of hemophilia
- Identify person at risk

THANKYOU

COMPLICATIONS

Liver abnormalities

Hematuria

Renal disease

HTN

Nursing diagnosis

- Risk for hemorrhage related to deficiency of coagulation factor
- Acute pain related to bleeding into the muscles and joint cavities
- Risk for trauma related to tooth extraction and injection
- Impaired physical mobility related to joint stiffness
- Risk for infection related to frequent blood transfusion

- Altered growth and development related to life threatening illness
- Fear and anxiety related to poor prognosis
- Knowledge deficit regarding disease condition and its management related to lack of information
- Altered family process related to repeated hospitalization.
- Altered parenting related to the cost therapy

PATHOPHYSIOLOGY

- An inherited deficiency of factor VIII or IX (antihemophilic factors)

CLASSIFICATION

- The normal level of factor VIII is 60 to 140%
- The normal levels of factor IX is 60-145%.
- A factor level of 30% or less of a normal level is diagnostic of hemophilia & may be classified as

Clinical severity	Factor VIII activity	Bleeding tendency
Severe	<2%	Spontaneous bleeding without trauma
Moderate	2%-49%	Bleeding with trauma
Mild	5-40%	Bleeding with severe trauma or surgery