Lecture 5 ---------- blood transfusion

Hemolytic Disease of the Newborn

Hemolytic disease of the newborn is also called **erythroblastosis fetalis**. This condition occurs when there is an incompatibility between the blood types of the mother and fetus.

What causes hemolytic disease of the newborn (HDN)?

Hemolytic Disease of the Newborn (HDN) and Rh Incompatibility

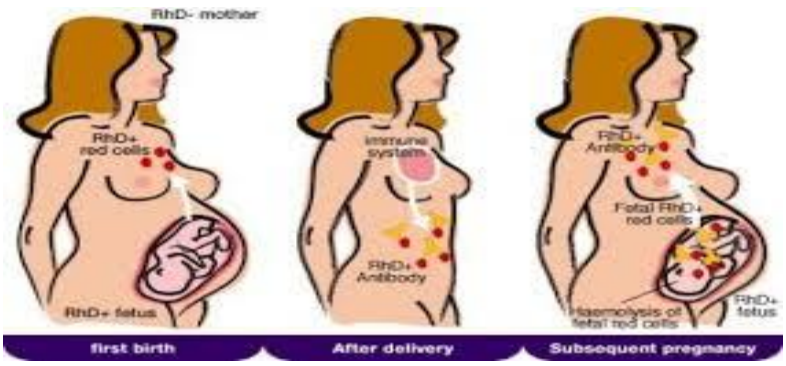
1. Cause: HDN most commonly occurs when an **Rh-negative mother** carries an **Rh-positive baby**. The mother's immune system recognizes the baby's Rh-positive red blood cells as **foreign**.

2. Immune Response:

- The mother's immune system produces **antibodies** against the Rh antigen to destroy the foreign red blood cells. Once these antibodies are formed, the mother becomes **Rh-sensitized**, meaning her immune system "remembers" the Rh antigen and can respond more strongly in future pregnancies.

3. First Pregnancy & Subsequent Pregnancies:

- During the first pregnancy, Rh sensitization **usually does not occur**, so the baby is typically unaffected. If the mother is Rh-sensitized and becomes pregnant with another **Rh-positive baby**, her antibodies can **cross the placenta**. These antibodies attack the baby's red blood cells, causing **hemolysis** (destruction of red blood cells).
- The destruction of red blood cells can lead to **anemia, jaundice, or more severe complications** in the newborn. This condition is called **Hemolytic Disease of the Newborn (HDN)**.



Complications of HDN:

Complications of hemolytic disease of the newborn can range from mild to severe.

1. Mild anemia, hyperbilirubinemia, and jaundice.
2. Severe anemia with enlargement of the liver and spleen. When these organs and the bone marrow cannot compensate for the fast destruction of red blood cells, severe anemia results and other organs are affected.
3. Hydrops (fluid throughout the body's tissues, including in the spaces containing the lungs, heart, and abdominal organs), which can lead to heart failure from too much fluid
4. Kernicterus is the most severe form of hyperbilirubinemia and results from the buildup of bilirubin in the brain. This can cause seizures, brain damage, deafness, and death.

What are the symptoms of hemolytic disease of the newborn?

1. A pale coloring may be evident, due to anemia.
2. Jaundice or yellow coloring of skin and eyes may be present. jaundice can develop quickly, usually within 24 to 36 hours.
3. The newborn may have an enlarged liver and spleen

Prevention of Hemolytic Disease of the Newborn (HDN)

1. Early Identification: HDN is highly preventable. **All pregnant women** are tested early in pregnancy for **Rh factor**. If the mother is **Rh-negative**, preventive measures are taken.

2. Rh Immunoglobulin (RhIg / RhoGAM): A specially developed blood product that prevents an Rh-negative mother's antibodies from reacting to Rh-positive red blood cells. Other Uses: Can also be used when Rh-negative individuals receive Rh-positive blood transfusions. Administration: Injected into a muscle (IM) or vein (IV).

3. Timing and Dosage:

- **After delivery:** Many Rh-negative mothers receive RhoGAM **within 72 hours** if the baby is **Rh-positive**.
- **No dose is needed** if the baby is **Rh-negative**.

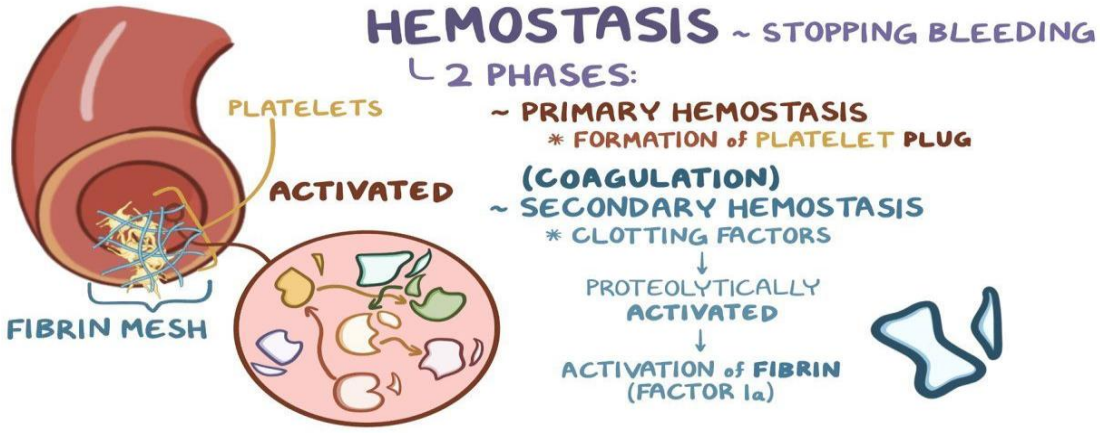
How is hemolytic disease of the newborn diagnosed?

Once a baby is born, diagnostic tests for HDN may include the following: Complete blood count and immature red blood cell (reticulocyte) count , Bilirubin level& Blood typing.

HEMEOSTASIS & BLEEDING DISORDER

Hemostasis is the rapid, localized, and carefully controlled process by which the body stops bleeding (arrests hemorrhage) from a damaged blood vessel, while maintaining blood in a fluid state within the rest of the circulatory system. It's a protective mechanism against excessive blood loss. **Hemorrhage** (excessive bleeding) and **Thrombosis** (excessive or inappropriate clotting). **Components of hemostasis :**

- **Blood Vessels** (Endothelium and smooth muscle)
- **Platelets** (Thrombocytes)
- **Coagulation Factors** (Plasma proteins)



Stages of Hemostasis : Hemostasis is the process the body uses to stop bleeding after blood vessel injury. It occurs in overlapping stages:

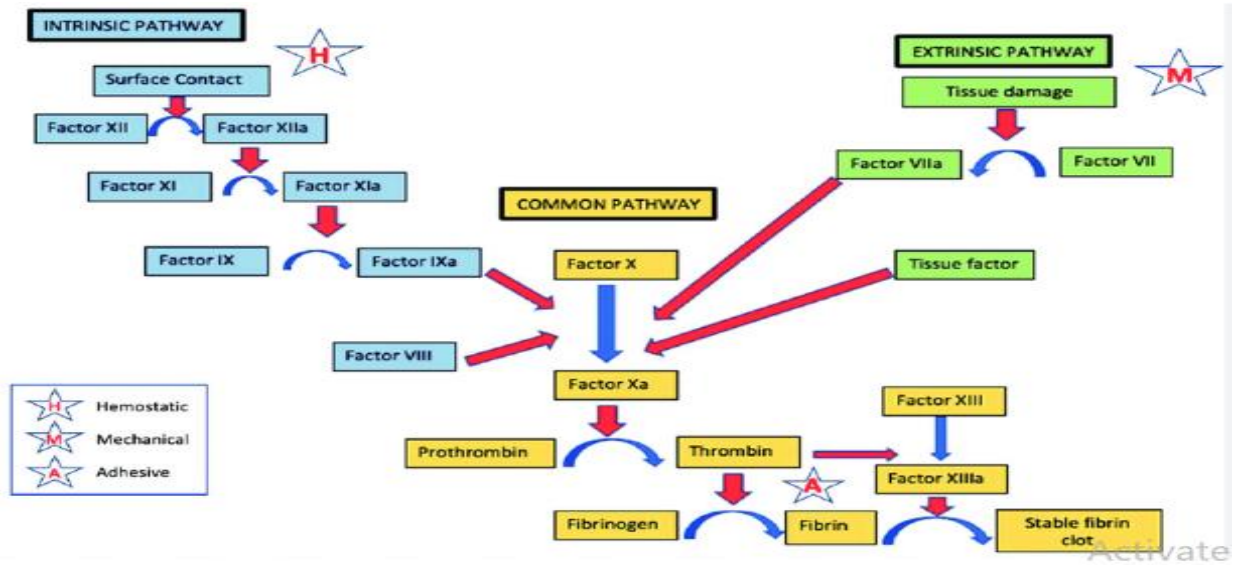
1. Primary Hemostasis – Formation of the Platelet Plug

Vascular spasm (vasoconstriction) is the very first response after a blood vessel is injured. The damaged vessel immediately constricts to reduce blood loss. This constriction occurs because the smooth muscle in the vessel wall is injured and due to the release of vasoconstrictor substances such as serotonin and thromboxane A_2 from platelets and endothelial cells. After this, platelet plug formation begins. Platelets first adhere to the exposed collagen at the site of injury, a process that depends mainly on von Willebrand factor, which helps anchor the platelets

to the damaged surface. Once they adhere, the platelets become activated, change shape, and release important factors from their granules, including Adenosine Diphosphate (ADP) and thromboxane A_2. These substances recruit and activate additional platelets. As more platelets are activated, they start to stick to each other, forming a temporary platelet plug that provides an initial seal to reduce bleeding.

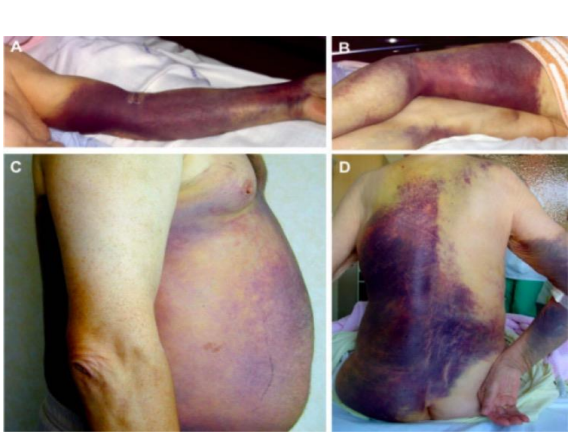
2. Secondary Hemostasis – Formation of the Fibrin Clot

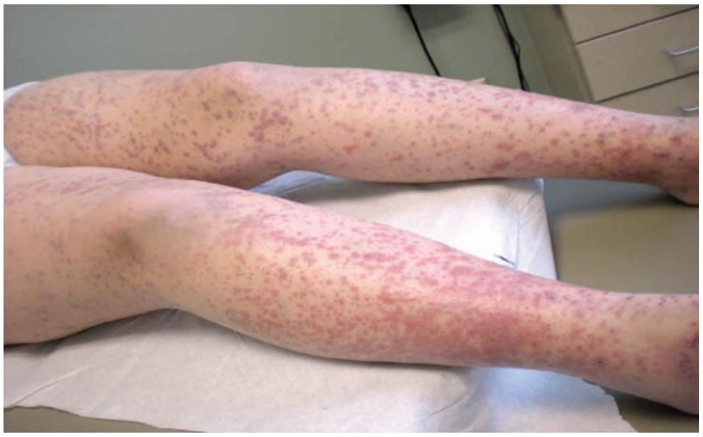
Secondary hemostasis involves the coagulation cascade, which converts soluble fibrinogen into insoluble fibrin to strengthen and stabilize the platelet plug. The cascade can be initiated by two pathways: **the extrinsic pathway**, triggered by external trauma and tissue release of Tissue Factor, activates Factor VII and serves as the primary initiator; **the intrinsic pathway** activated by vessel damage and exposure of blood to collagen, involves Factors XII, XI, IX, and VIII and mainly amplifies the coagulation process. Both pathways converge at Factor X in the common pathway, forming the prothrombin activator complex that converts prothrombin into thrombin. Thrombin then converts fibrinogen into fibrin, and Factor XIIIa cross-links the fibrin strands to produce a firm, stable clot. Fibrinolysis (Clot Dissolution - Tertiary Hemostasis)A mechanism to prevent the clot from growing too large and to dissolve it once the vessel is repaired. Plasminogen is converted to the active enzyme Plasmin, which then breaks down the fibrin mesh into Fibrin Degradation Products (FDPs).



Bleeding disorders

Bleeding disorders, or hypocoagulable states, occur when one or more components of the hemostatic system fail, leading to problems such as easy bruising, prolonged bleeding, or spontaneous hemorrhage. Disorders of platelets or blood vessels affect primary hemostasis and usually cause immediate, superficial bleeding such as petechiae and mucosal bleeding.





Specific bleeding disorders include:

- Acquired platelet function defects
- Congenital platelet function defects
- Disseminated intravascular coagulation (DIC)
- Coagulation Factors deficiency
- Hemophilia A, B & C
- Idiopathic thrombocytopenic purpura (ITP)
- Von Willebrand's disease (types I, II, and III)

- **Thrombocytopenia** refers to a reduced platelet count, which may result from decreased production in the bone marrow, increased destruction as seen in immune thrombocytopenic purpura, or increased consumption. Platelet function defects involve abnormal platelet activity even when the platelet count is normal.
- **Von Willebrand disease**, the most common inherited bleeding disorder, results from a deficiency or abnormality of von Willebrand factor, impairing platelet adhesion and lowering Factor VIII levels. Some bleeding disorders also arise from abnormalities in the vessel wall, although these are less common. Coagulation factor disorders involve defects in secondary hemostasis and typically present with delayed, deep bleeding into joints and muscles.
- **Hemophilia A** is caused by a deficiency of Factor VIII and is inherited in an X-linked recessive pattern, primarily affecting males; it is characterized by recurrent joint bleeding, muscle hematomas, and prolonged bleeding after trauma or surgery.
- **Hemophilia B**, caused by a deficiency of Factor IX, has the same inheritance pattern and similar clinical features.
- **Severe liver disease** can produce multiple clotting factor deficiencies because the liver synthesizes most coagulation proteins. Vitamin K deficiency also impairs coagulation, since Vitamin K is required for the synthesis of Factors II, VII, IX, and X; warfarin works by inhibiting Vitamin K activity.

Symptoms: common Symptoms may include any of the following:

- Bleeding into joints or muscles
- Heavy bleeding after injury
- Excessive bleeding with surgical procedures

Tests for bleeding disorder include:

- Diagnosis of bleeding disorders relies on both screening and specific tests. Platelet count and bleeding time help evaluate primary hemostasis, although bleeding time is used less frequently today. Prothrombin time assesses the extrinsic and common pathways, while activated partial thromboplastin time evaluates the intrinsic and common pathways. When needed, individual factor assays can be performed to measure the level or activity of specific clotting factors, such as Factor VIII in suspected Hemophilia A.

- Complete blood count (CBC)
- Bleeding time
- Partial thromboplastin time (PTT) : important for hemophilia and monitoring
- Platelet aggregation test
- Prothrombin time (PT) :for **Factor VII ,X,V Factor II (Prothrombin) Factor I (Fibrinogen):**
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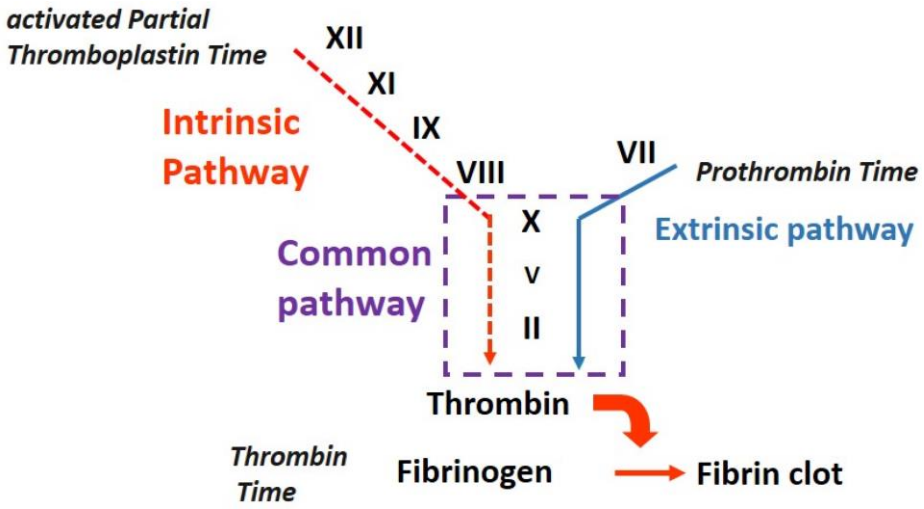

Treatment : Treatment depends on the type of disorder. It may include:

- Clotting factor replacement
- Fresh frozen plasma transfusion
- Platelet transfusion
- Other treatments