Hydrops Fetalis – Dr. Haidar

It is a condition occur in fetus and characterized by anasarca, ascites, pleural and pericardial effusions, hepatosplenomegaly, hypoalbuminemia and congestive heart failure.

Causes:
1. **Immune hydrops:** like in ABO incompatibility, Rh and other incompatibilities with intrauterine hemolysis of the fetal erythrocytes by maternal immunoglobulin IgG sensitized antibodies crossing the placenta.
2. **Non-immune hydrops:** like in supraventricular tachycardia, fetal anemia (resulting from bone marrow suppression), non-immune hemolysis, twin-twin transfusion syndrome, severe congenital malformations, congenital infections and inborn error of metabolism.

Haematological Disorders

Anaemia

- Anemia in newborn is defined as a hemocrit less than 40% (normally hemocrit at term gestation is 50-55%).
- **Causes:**
  - The principle causes of anemia in the newborn can be divided into those associated with acute blood loss, those associated with chronic blood loss and those associated with impaired red cell production.
  - **Acute blood loss:** like placenta previa, placental abruption, feto-maternal transfusion, feto-placental transfusion, cord rupture, and internal hemorrhage.
  - **Chronic blood loss:** like hemolytic disease, twin-twin transfusion, feto-maternal transfusion, chronic phlebotomy.
  - **Impaired red blood cell production:** like congenital hypoplastic anemia (Diamond Blackfan syndrome).

- **Hemoglobin synthesis:** Embryonic hematopoiesis begins by the 20th day of gestation and is evidenced as blood islands in yolk sac. In mid gestation, erythropoiesis occurs in the liver and spleen; the bone marrow becomes the predominant site in the last trimester. Hb concentration increases from 8-10 g/dl at 12 wk to 16.5-18.5 g/dl at 40 wk.
- In the last two trimesters of the pregnancy produces fetal hemoglobin (Hb F) which composed of two alpha chains and two gamma chains. Prior to term, the infant begins to synthesize beta- Hb chains; thus the term infant should have some adult Hb (2 alpha and 2 beta chains). Fetal Hb represents 60-90% of Hb at birth, and the level decline to adult levels of less than 5% by 4 mo of age.
- The blood volume of the term infant is approximately 85 ml/kg.
- The placenta and umbilical vessels contain approximately 20-30 ml/kg of additional blood that, if clamping or milking of the umbilical cord or delayed clamping of the cord may increase the risk of polycythemia, on the other hand early clamping of the umbilical cord may result in anemia.

Diagnosis of neonatal anemia:

The specific cause of the anemia is established on the basis of information collected from the following sources:

1- History
2- C.B.C. with peripheral smear and reticulocyte count
3- Evaluation of maternal and infant blood or Rh or ABO incompatibility
4- Coombs test
5- Other tests like Kleihauer test (to identify & quantify fetal red blood cells), Hb electrophoresis, & G6PD evaluation.

Therapy:

- Acute blood loss should be treated rapidly. Therapy includes restoration of blood volume and red blood cell mass and elimination of the cause of blood loss.
- Chronic blood loss therapy varies depending on the clinical condition & cause of blood loss & may consist transfusion of packed red cells, partial exchange transfusion with packed red cells, iron therapy or no intervention.
Hemolytic disease of the newborn (erythroblastosis fetalis):

- Usually results from blood group incompatibility between the mother and the fetus. Hemolysis occurs when maternal Ab to a particular blood group Ag cross the placenta and bind to fetal blood cells, which are then destroyed in the spleen.
  1. The most commonly involved Ag is Rh (D) Ag.
  2. ABO blood group Ag are less commonly involved.
  3. Rarely, hemolytic disease of the newborn is caused by other blood group incompatibilities (e.g. c,E,Kell), or G6PD deficiency or vitamin E deficiency.
  4. If anemia severe, hydrops fetalis may developed.

- **Therapy:** Therapy is indicated when Hb and hematocrit are low enough to compromise the oxygen–carrying capacity of the blood, which can cause congestive heart failure, respiratory distress, acidosis, poor perfusion and hypotension. The blood volume usually is normal. Therefore, the anemia is corrected by performing a partial exchange transfusion with packed red blood cells.

- **Prevention:** Prevention of sensitization of the mother carrying an Rh–positive fetus is possible by treating her during pregnancy (after 28 wk) and within 72 hr after birth with anti-Rh- positive immune globulin.

Hemorrhagic disease of the newborn:

- A disease primarily of breast fed infants, occurs at 2–10 days of life in those who do not receive prophylactic vitamin K on the first day of life (1-2% of all newborn may be affected)

- **Clinical features:** bleeding usually occur from umbilical cord, circumcision site, intestines, scalp, mucosa and skin, and the most dangerous fatal hemorrhage is the internal hemorrhage like intracranial bleeding.

- If hemorrhage occur in the first day of life so most probably resulting from a deficiency of the vitamin K dependent factors, often associated with administration of drugs to the mother during pregnancy that affecting the vitamin K metabolism like Warfarin or Phenobarbital and Phenytoin.

- **Prevention:** by administration of vitamin K to all infants at birth, 1mg of vitamin K given I.M. will prevent bleeding

- **Treatment:** treatment of bleeding resulting from vitamin K deficiency requires I.V. administration of vitamin K. If bleeding is severe and life threatening hemorrhage is present, so fresh frozen plasma should be given.

Polycythemia (hyper-viscosity syndrome):

- **Definition:** Polycythemia is a central venous hematocrit greater than 65%. It is usually occur in 2-4 % of infants born at sea level.

- **Predisposing factors:** infants at special risk for polycythemia are:
  1. Term , post term and SGA infants
  2. Infants of diabetic mothers
  3. Infants with delayed cord clamping
  4. Neonatal hyperthyroidism
  5. Adrenogenital syndrome
  6. Trisomy 21 = Down syndrome
  7. Twin – twin transfusion syndrome
  8. Beckwith-Weidemann syndrome
  9. polycythemia may reflect a prolonged period of fetal hypoxemia

- **Clinical features:** Manifestation include plethora, poor perfusion, cyanosis, poor feeding, respiratory distress, lethargy, jitteriness, seizures, renal vein thrombosis and metabolic acidosis and there will be increased risk for necrotizing enterocolitis, hypoglycemia, thrombocytopenia and hyperbilirubinemia.

- **Complications:** In addition to the previous features, complications of polycythemia are mainly related to the primary condition associated with polycythemia like post-maturity, SGA,...

- **Management:** Treatment includes partial exchange transfusion, in which blood is removed and replaced by the same volume of plasma substitute (like normal saline, plasma, and albumin) in a stepwise manner.
  - Volume to exchange (ml) = \( \frac{\text{Blood Volume (Observed Hct – Desired Hct)}}{\text{Observed Hct}} \)
  - The desired Hct is 50% and the blood volume is 85 ml/kg