Pediatrics – Dr. Haidar – Lecture 4 - Prevention of Rh incompatibility hemolytic disease

Prevention of sensitization of the mother carrying an Rh +ve fetus is possible by treating her during gestation (after 28 wk) and within 72 hr after birth with anti Rh positive immunoglobulin (RhoGAM) 300 Mg, this based on the ability of this amount of anti Rh positive Ab to bind all the possible fetal Rh positive erythrocytes entering the maternal circulation during the fetal to maternal transfusion. RhoGAM is effective only in preventing sensitization to the D antigen.

Treatment:
There are many ways for treating hyperbilirubinemia, but the main ways are the following:

Phototherapy:-

Is an effective and relatively safe method for reducing indirect bilirubin levels, particularly when initiated before serum bilirubin increases levels associated with Kernicterus. In term infants, phototherapy is begun when indirect bilirubin levels are between 15 and 18 mg/dL, while in preterm infants it is initiated at lower levels to avoid bilirubin reaching the high concentration requiring exchange transfusion.

Bilirubin absorbs light maximally in the blue range (420-470 nm), broad spectrum white, blue, special narrow spectrum (super) blue, and less often, green lights have been effective in reducing bilirubin levels. Bilirubin in skin absorbs light energy which by photo–isomerization converts the toxic native lipid soluble unconjugated bilirubin into water soluble unconjugated bilirubin which is excreted in bile without need for conjugation.

Phototherapy also converts native bilirubin by an irreversible reaction to the structural isomer lumirubin which is excreted by the kidneys in the unconjugated state. Phototherapy has decreased the need for the exchange transfusion in term and preterm infants, but should not be used as substitute for exchange transfusion.

In preterm infants without significant hemolysis, serum bilirubin usually decline 1-3 mg/dL after 12-24 hr of phototherapy. The therapeutic effects depends on the light energy emitted in the effective range of wavelengths, the distance between the light and the infants and the amount of skin exposed, as well as the rate of hemolysis.

Phototherapy includes special blue fluorescent tubes placing the lumps within 15-20 cm from the infant and placing a fiberoptic phototherapy blanket under the infants back to increase the exposed surface area.

The infant’s eyes should be closed and adequately covered to prevent exposure to light which may be toxic to retina. Also gonads should be covered as well.

Phototherapy is applied continuously, and the infant is turned frequently for maximal skin exposure, it should be continued as soon as the indirect bilirubin concentration has been reduced to levels considered safe in view of the infant’s age and condition. Some time it used in alternative method (on and off 6 hr alternative).

Complications:- increased in insensible water loss, diarrhea, dehydration, maculopapular skin rash, lethargy, masking of cyanosis, pyridoxine deficiency, nasal obstruction due to eye pads, and potentially retinal damage, some time skin bronzing (bronz baby) due to direct hyperbilirubinemia.

Exchange Transfusion:-

Usually is indicated for infants with dangerously high direct bilirubin levels who are at risk for kernicterus (indirect hyperbilirubinemia >20-25 mg/dL). Asymptomatic infants with
physiologic or breast milk jaundice may not require exchange transfusion unless indirect bilirubin level approaches 25 mg /dl or more.

Blood for exchange transfusion should be as fresh as possible, heparin or citrate may be used as anticoagulant. If the blood is obtained before delivery, it should be taken from type O Rh–ve donor with lower titer of anti-A and anti-B Ab and should be compatible with the mother serum by the indirect Coombs test. After delivery, blood should be obtained from an Rh–ve donor whose cells are compatible with both the infant’s and the mother’s serum; when possible type O–ve donor cells are generally used. But cells of the infant’s ABO blood type may be used when the mother has the same type.

A complete cross match, including indirect Coombs test should be performed before the second and subsequent transfusions. Blood should be warmed gradually and maintain temp. between 35-37°C throughout the exchange transfusion. It should be kept well mixed by gentle squeezing or agitation of the bag to avoid sedimentation.

with a strict aseptic technique, the umbilical vein is cannulated with a polyvinyl catheter to a distance no greater than 7 cm in a full term infant. When free flow of blood is obtained, the catheter is usually in a large hepatic vein or inferior vena cava.

The exchange should be carried out over a 45-60 min (or 45-90 min in ill infant).

aspiration of 20 ml of infant blood alternating with infusion of 20 ml of donor blood. Smaller amounts (5-10 ml) may be indicated for sick and premature infants.

The goal should be an isovolumetric exchange of approximately two blood volumes of the infant (2 x 85 ml/kg), that mean amount require for exchange equal to = body weight x 2 x 85 ml.

Calcium should be given during the procedure, because hypocalcemia is one of the commonest complication of this procedure (due to presence of citrate in blood bag)

during the procedure all drugs and tools for resuscitation should be available (like adrenaline, Na Bicarbonate, atropine, diazepam, i.v. fluid ...etc) in order to treat the possible immediate complications that may occur during the procedure.

Complications :-

Can be divide into :

1) complications resulting from blood transfusion like transfusion reaction, metabolic instability, infections, etc.

2) complications resulting from catheter like vessel perforation and hemorrhage.

3) complications resulting from the procedure like hypotension, necrotizing enterocolitis.

or can be classified into early and late complications:

Early like: transf. reaction, metabolic instability, vessels perforation, bleeding, hypotension, hypocalcemia and hypothermia.

Late complications like: infections (H.C. H.B, H.I.V.), N.E.C., and Graft versus host disease.

Kernicterus (bilirubin encephalopathy) :-

Lipid soluble, unconjugated, indirect bilirubin fraction is toxic to the developing central nervous system, especially when indirect bilirubin concentrations are high and exceed the binding capacity of albumin.
kernicterus results when indirect bilirubin is deposited in brain cells, disturbing neuronal metabolism and function, especially in basal ganglia. Indirect bilirubin may cross the blood brain barrier because of its lipid solubility.

Kernicterus usually is noted at an excessively high bilirubin level for gestational age (20-25 mg/dL for full term infants).

*Kernicterus may be noted at bilirubin levels below 20 mg/dL like in the followings:*

1- sepsis 2- meningitis 3- hemolysis 4- asphyxia 5- hypoxia 6- hypothermia 7- hypoglycemia 8- bilirubin displacing drugs 9- prematurity and very low birth infant may develop kernicterus when their bilirubin levels are less than 10 mg/dL.

**Clinical manifestations:**

*Early manifestations like:* lethargy, hypotonia, poor moro reflex and poor feeding

also high pitched crying and vomiting may be present.

*Late manifestations like:* bulging fontanel, opisthotonic posturing, pulmonary hemorrhage, fever, hypertonicity, and seizures.

*Late sequelae or complications:* noticed usually in survived neonates and include nerve deafness, choreoathetoid C.P., mental retardation, enamel dysplasia and discoloration of teeth (yellow).

**Prognosis:** quietly poor prognosis.

**Prevention:** by avoiding excessively high indirect bilirubin levels and by avoiding conditions that may displace bilirubin from albumin and also by treatment of neonatal jaundice by phototherapy or exchange transfusion.

**Neonatal Direct Hyperbilirubinemia:**

Most common causes in neonates like Biliary atresia, choledocal cyst, neonatal sepsis, neonatal hepatitis, Galactosemia, alpha-1-antitrypsin deficiency and congenital infections (TORCH).

sites for taking blood sample in neonate ---->