A Newborn with Persistent Hyperbilirubinemia

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Presentation

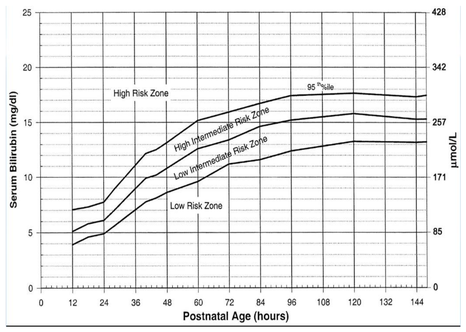
- ❑ Male, African American infant weighing 2999 g, born via normal spontaneous vaginal delivery at 37 weeks and 4 days to a 27 yo G3P1A1 mother.
- ❑ Apgar scores were 9/9
- ❑ Mother tested positive for Group B Strep and was adequately treated with penicillin.
- ❑ Mother was an alpha thalassemia carrier, but father of the baby refused testing.
- ❑ Mother was B positive, infant was O positive and the direct antiglobulin test was negative.
- ❑ Remainder of prenatal history was unremarkable.

Physical Exam

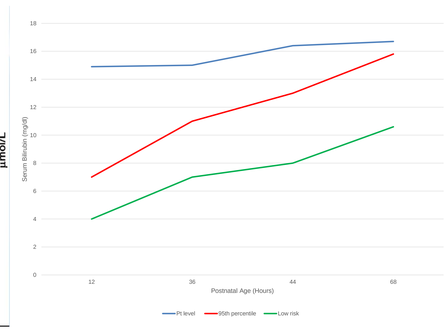
- ❑ Remarkable for obvious jaundice and a distended abdomen.
- ❑ Baby passed 3 substantial meconium plugs.
- ❑ Transcutaneous bilirubin level 15.1 at 24 hours of life.
- ❑ Confirmed with a serum bilirubin of 14.9.

Course of Action

- **24 hours of life**: Serum Bilirubin 14.9; Phototherapy Initiated
- **36 hours of life**: Serum Bilirubin 15.0; Double Phototherapy Initiated
- **44 Hours of life**: Serum Bilirubin 16.4; CBC with reticulocyte count & G6PD ordered
- **NICU transfer**: •Triple Phototherapy & IV Fluids without success
- **68 hours of life**: Serum Bilirubin 16.7; Reticulocyte count, Hemoglobin, G6PD , T4, TSH all WNL; Blood cultures negative

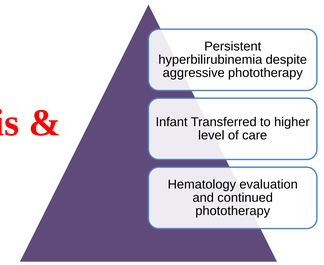


Nomogram for Serum Bilirubin by Postnatal Age (Hours)



Graph showing patient serum bilirubin levels compared with 95%ile and low risk

Final Diagnosis & Outcome



- ❑ Elevated mean corpuscular hemoglobin concentration (MCHC) at 36.8 and a low mean corpuscular volume (MCV) at 77.4 were noted which is consistent with a diagnosis of Hereditary Spherocytosis.
- ❑ The infant responded well to aggressive IV hydration and phototherapy.
- ❑ Patient was discharged home with hematology follow-up 2 weeks after discharge.

Risk Factors for Hyperbilirubinemia

ABO incompatibility

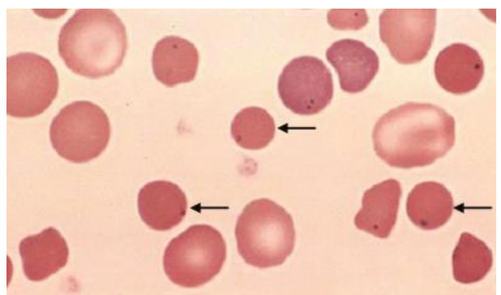
Prematurity with gestational age of 35-36 weeks

Cephalohematoma or bruising

Breastfeeding

Differential Diagnoses

- Glucose-6-phosphate dehydrogenase
- Hereditary Spherocytosis
- Thalassemia
- Meconium Plug Syndrome



Discussion

- Hereditary Spherocytosis (HS) is the most common cause of nonimmune hemolytic anemia due to an inherited red blood cell membrane disorder leading to mild to severe neonatal jaundice affecting 1 in 2,000-5,000 people.
- HS is characterized by spherical red blood cells in the peripheral blood. Infants with HS will present a variety of ways from asymptomatic to severe fetal anemia with hydrops fetalis.
- In the neonatal period, jaundice is the most common presentation many times without hepatosplenomegaly or anemia. The peripheral smear of the neonate may also not show any typical spherocytes.
- The mean corpuscular hemoglobin concentration (MCHC) and mean corpuscular volume (MCV) are most helpful when suspecting HS. An elevated MCHC and a low MCV, and a MHCH/MCV ratio >0.36 are used to diagnose infants with HS.
- Rarely other tests are used to confirm the diagnosis including an epithelial membrane antigen and osmotic fragility test.
- Infants with suspected HS are treated with phototherapy, exchange transfusions to avoid kernicterus if needed, and packed red blood cell transfusions for symptomatic anemia. These patients should be followed closely by hematology.

References:

Pan, D H & Rivas, T. Jaundice: Newborn to Age 2 months. *Pediatrics in Review* 2017; 38: 499.

Mahajan V & Jain S K. Hereditary Spherocytosis. *NeoReviews* 2016; 17: e697.