

## Hemolytic Anemia In A Neonate: A Diagnostic Dilemma

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### Dear Sir,

Medical literature is replete with cases of hemolytic anemias in newborn. Spectrum of hemolytic diseases of the newborn (HDN) has changed beyond recognition in the past few decades. Hemolysis was almost synonymous with Rh D allo-immunization a decade ago.<sup>[1]</sup> With better understanding of the disease and great improvements in antenatal diagnosis, overall morbidity and mortality has come down exorbitantly. However, neonatal hemolysis still remains a huge diagnostic dilemma for clinicians and pathologists alike.

Here, we report a case of 5 days old neonate who presented with jaundice and severe hemolytic anemia requiring intensive phototherapy and blood transfusions. With an exhaustive workup of the case, the cause still remains intangible.

A 5 days old male baby was brought with complaints of icterus since 2 days. Baby was born to 30 year old second gravida at term by vaginal delivery with weight of 2.2 kg with features of IUGR. Mother's blood group was B positive. No history of lethargy, vomiting, or respiratory distress. There was no history of any maternal illness complicating pregnancy. No family history of frequent blood transfusions or blood dyscrasias.

On examination, the baby weighed 2.15 kg. He had icterus till forearms and legs with palms and soles being spared. There were no features of lethargy, high pitched cry or abnormal posturing. No cephalhematoma, or hepatosplenomegaly.

Initial laboratory evaluation revealed a Hemoglobin of 13 gm/dl with serum bilirubin being 21 mg/dl (conjugated being 0.9mg/dl). Further sample was sent for DCT, which was negative with baby's blood group being B positive. Baby was put on intensive phototherapy with consideration of IVIG in case of rising trend of bilirubin nearing exchange transfusion range. With 48 hours of intensive phototherapy serum bilirubin values showed a decreasing trend with value reaching 14.5 mg/dl for which phototherapy was

stopped. However the bilirubin values again rose to 23 mg/dl within 24 hours of stoppage of phototherapy. Follow up hematological investigation revealed a severe fall in Hemoglobin (Graph 1) with peripheral blood smear showing features of hemolysis with normal RBC indices (MCHC 33 gm/dl). Osmotic fragility test and G6PD test were normal. HPLC of the parents showed no features of hemoglobinopathy and G6PD test also being normal. Baby was transfused with PRBC at HCT below 21% and phototherapy was continued for a total of 7 days. A gradual fall in bilirubin levels was seen (Graph 2) to reach a level of 12 mg/dl.

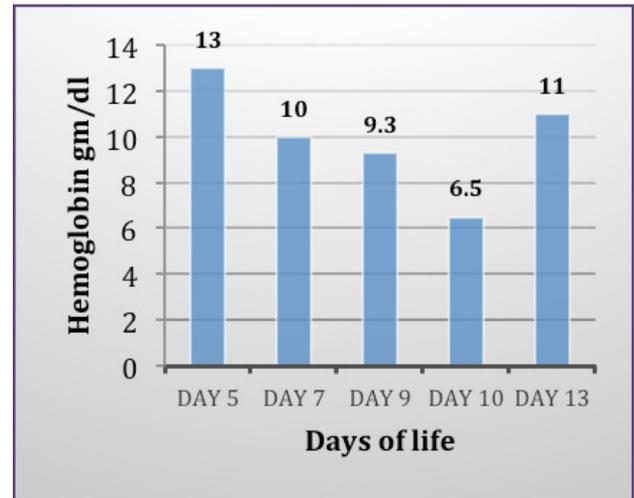
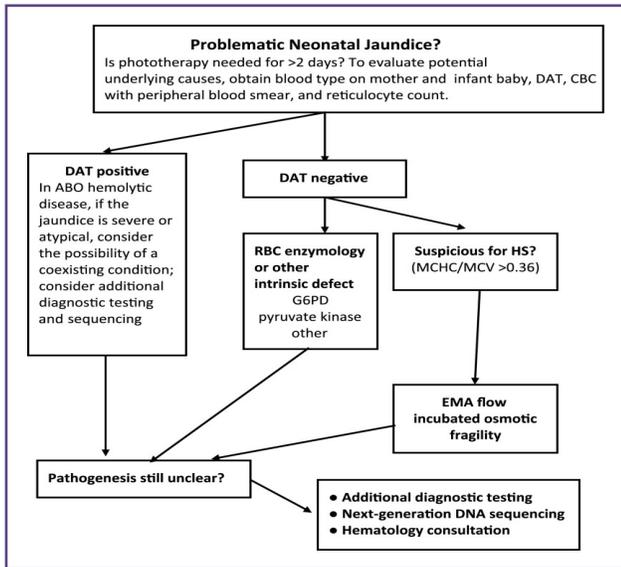
Repeat G6PD test was done which was normal along with normal 5 EMA studies, RBCs pyruvate kinase enzyme was also found to be normal. He was again transfused with PRBC in view of falling hemoglobin and HCT after 1 month of initial transfusion.

Presently the baby is asymptomatic and thriving well with Hb nadir of 9.3 gm/dl with no evidence of hemolysis.

Jaundice in neonates is one of the commonest presentation that pediatricians manage on daily basis. To make matters worse is the presence of severe ongoing hemolysis and need for repeated blood transfusions. The AAP has laid down algorithmic approach in such cases (Figure 2).<sup>[2]</sup> With DCT being negative the list of probable causes narrow down to a few conditions, of which HS and G6PD deficiency are the commonest. However, both of these tests done repeatedly were negative in our case. Next generation DNA sequencing would be next step in this case but with resource limited setting like ours, the diagnosis still remains ambiguous.

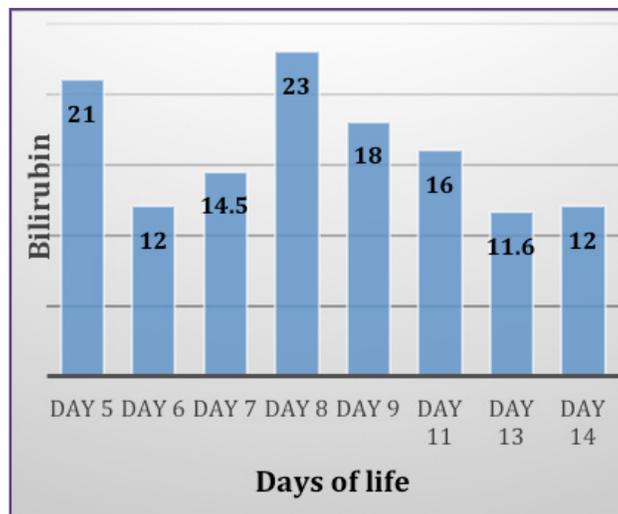
### Reference

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Graph 1: Trends in fall and rise of Hemoglobin

Fig. 1: Algorithmic approach to DCT negative hemolytic anemia [2].



Graph 2: Bilirubin Trend

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