

An Approach to Conjugated Hyperbilirubinemia in Neonates



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INTRODUCTION

Cholestatic jaundice affects around **1 in 2,500** infants, highlighting the need for timely diagnosis via efficient neonatal screening processes. Cutaneous and scleral icterus is a common finding in the first 2 weeks after birth and is regarded as physiological jaundice. Increased conjugated bilirubin due to reduced bile flow is an indication of serious **hepatobiliary/metabolic dysfunction**. Early diagnosis of the same is essential for preventing progression into liver failure or serious extrahepatic complications. We present below a **case of a 24-day-old neonate** whose parents gave a history of **conjugated hyperbilirubinemia**.

CASE REPORT

A **24-day-old neonate** was brought to neonatologist with **persistent yellowish discoloration of sclera and skin and pale stools**.

As per history, the neonate was born to non-consanguineous marriage and had a Full Term Normal Vaginal Delivery (FTNVD). The antenatal, perinatal, and postnatal history was uneventful. Breast feeding was initiated immediately after birth. Yellowish discoloration was seen from day 3, however it was dismissed as physiological jaundice, and the neonate was discharged. **About three weeks after discharge, the baby was brought back to the hospital with persistence of icterus.**

Examination:

- The neonate appeared **lethargic** and had a **weak cry**.
- Was **not feeding properly**
- Yellowish discoloration** of the sclera and skin
- Distension of abdomen** with **hepatomegaly**

Lab investigations:

Parameter	Value	Age specific biological reference interval
Hemoglobin (Hb)	11 g/dL	13.4-19.9 g/dL
Total Count (TC)	14000/cumm	9000 – 30,000/ cu mm
Total Bilirubin (TB)	9 mg/dL	0.3-1 mg/dL
Direct Bilirubin (DB)	6 mg/dL	0.1-0.3 mg/dL
AST	154 U/L	<40 U/L
ALT	187 U/L	<40 U/L
ALP	357 U/L	90 – 180 U/L
GGT	165 U/L	<50 U/L

References:

- Feldman AG, Sokol RJ. Neonatal Cholestasis: Updates on Diagnostics, Therapeutics, and Prevention. NeoReviews. 2021 Dec 1;22(12):e819–36.
- Kelly DA, Stanton A. Jaundice in babies: implications for community screening for biliary atresia. BMJ. 1995 May 6;310(6988):1172–3.

DISCUSSION

The patient has neonatal cholestasis as there is persistence of icterus after 2 weeks of birth and pale stools has been reported. Laboratory investigations also show **elevated direct and total bilirubin** as well as **liver enzymes**. Distended abdomen and hepatomegaly also may be an indicator of an obstruction of bile flow.

Considering **primary biliary atresia** as a cause, wherein the ducts (inside/outside liver) carrying bile from liver to gall bladder fail to develop due to a congenital defect, hepatomegaly and distended abdomen is well explainable due to back flow of the bile juice. Consequently, the conjugated bilirubin is not converted to stercobilinogen in the intestine that leads to **pale colored stools**. The conjugated bilirubin even increases in the blood and deposits on sclera and skin.

MANAGEMENT

Surgical management was carried out. The surgery performed was the **Kasai portoenterostomy**. A Roux loop was created from the proximal jejunum and anastomosed with the cut surface at the level of porta hepatis.

CONCLUSION

This case enhances the available knowledge regarding hyperbilirubinemia in neonates i.e. **conjugated hyperbilirubinemia** is always **pathological in neonates** and mandates prompt investigation into identifying the etiology.