

## PROLONGED JAUNDICE IN THE NEWBORN

### Definition

Visible jaundice (or serum bilirubin, SB >85  $\mu\text{mol/L}$ ) that persists beyond 14 days of life in a term infant or 21 days in a preterm infant.

**Table 1. Causes of prolonged jaundice**

Unconjugated Hyperbilirubinaemia	Conjugated Hyperbilirubinaemia
septicaemia or urinary tract infection (UTI)	biliary tree abnormalities
breast milk Jaundice	biliary atresia - extra, intra-hepatic
hypothyroidism	choledochal cyst
haemolysis	paucity of bile ducts
G6PD deficiency	- Alagille syndrome, non-syndromic
congenital spherocytosis	idiopathic neonatal hepatitis syndrome
galactosaemia	septicaemia or UTI
Gilbert syndrome	congenital infection (TORCHES)
	metabolic disorders
	citrin deficiency
	galactosaemia
	progressive familial intrahepatic cholestasis (PFIC)
	alpha-1 antitrypsin deficiency
	Total Parenteral Nutrition

### If unconjugated hyperbilirubinaemia:

- admit if infant is unwell. Otherwise follow-up until jaundice resolves.
- important investigations are; Thyroid Function Tests, Urine FEME, C&S and reducing sugar, and FBC, reticulocyte count & Peripheral Blood Film.
- exclude UTI and hypothyroidism.
- congenital hypothyroidism is a neonatal emergency. Check Screening TSH result if done at birth. (*see chapter on Congenital Hypothyroidism*)
- where indicated, investigate for galactosaemia
- breast milk Jaundice is a diagnosis of exclusion. Child must be well, have appropriate weight gain, feeds well with yellow stools. Management is to continue breastfeeding.

### If conjugated hyperbilirubinaemia:

(conjugated bilirubin > 2mg/dL or > 15% of total bilirubin)

- investigate for **biliary atresia** and neonatal hepatitis syndrome.

- admit and observe colour of stool for 3 consecutive days.
- further investigations: LFT, Hep B and C status, TORCHES, VDRL tests, alpha-1 antitrypsin and a metabolic (IEM) screen. ( $\gamma\text{GT}$ , GALT assay, Tandem Mass Spectrometry /IEM screen,  $\pm$  urine organic acids & plasma amino acid [refer chapter on IEM])
- consider Alagille syndrome
- **suspect biliary atresia if the stool is pale over 3 consecutive days.**

Refer paediatric surgery and plan for:

- *Ultrasound of the hepatobiliary system*
  - preferably done after 4 hours of fasting
  - dilated intrahepatic bile ducts and an absent gall bladder
  - raises the suspicion of extra hepatic biliary atresia

- HIDA Scan
  - increased accuracy of scan if oral phenobarbitone 5mg/kg given for 5 days, prior to the scan.
  - low uptake with normal excretion: neonatal hepatitis syndrome.
  - normal uptake with absent excretion: extrahepatic biliary atresia
- liver biopsy as indicated
  - biliary atresia can be confirmed in 85% by biopsy
  - ensure PT and aPTT normal before biopsy. Give Vitamin K 1 mg IV, if needed
  - platelet count  $\geq 40,000 /\text{mm}^3$  before biopsy
- operative cholangiogram followed by definitive surgery if necessary.
  - this is now the investigation of choice in most centres.

### **Biliary atresia**

- biliary atresia can be treated successfully by the Kasai procedure.
- this procedure must be performed within the first 2 months of life.
- with early diagnosis and biliary drainage through a Kasai procedure before 60 days of age, successful long-term biliary drainage is achieved in >80% of children. In later surgery good bile flow is achieved only in 20-30%.
- liver transplantation is indicated later if there is failure to achieve or maintain bile drainage

### **Neonatal Hepatitis Syndrome**

- follow up with LFT fortnightly. Watch out for liver failure and bleeding tendency (vitamin K deficiency).
- repeat Hepatitis B & C screening at 6 weeks
- most infants with idiopathic neonatal hepatitis make a complete recovery