

METABOLIC MANAGEMENT

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# **JAUNDICE (HYPERBILIRUBINAEMIA)**

Jaundice is usually a benign common condition in the newborn affecting 50% of term infants and at least 80% of those born before term. Management is aimed at risk assessment, recognition and appropriate treatment of hyperbilirubinaemia to prevent the development of severe hyperbilirubinaemia and the possibility of bilirubin encephalopathy.

# **INVESTIGATE THE FOLLOWING JAUNDICE**

## 1. APPEARING IN THE FIRST 24 HOURS OF LIFE

This is always important and must be investigated. It will most likely be due to haemolytic disease either associated with ABO incompatibility, rhesus isoimmunisation or due to one of the other rare antigens. If the infant shows evidence of skin haemorrhages such as petechiae, then a non-bacterial transplacental infection such as cytomegalovirus, toxoplasmosis, herpes or rubella is a possibility. Syphilis should also be considered.

## 2. OCCURRING AFTER THE FIRST DAY

Jaundice occurring on the second or third day of life is most likely to be due to physiological jaundice of the newborn. However, if the infant appears sick in any way, then other causes must be considered. Physiological jaundice is a diagnosis only arrived at by exclusion of more serious conditions.

## 3. OCCURRING BEYOND THE FOURTH OR FIFTH DAY OF LIFE

Generally this is not due to haemolytic disease so one must be on the lookout for bacterial infection particularly of the urinary tract or septicaemia. Again prenatally acquired infections should be considered. Jaundice due to drug interference is a possibility, and in infants of Asian or Mediterranean parents, glucose 6-phosphate dehydrogenase deficiency should be considered.

## 4. PERSISTING BEYOND THE FIRST TWO WEEKS OF LIFE

If bilirubin is mainly in the unconjugated form and the above mentioned conditions have been excluded then breast milk jaundice, hypothyroidism, galactosaemia and some of the other less common causes of jaundice should be considered.

### 5. CONJUGATED HYPERBILIRUBINAEMIA

If the total bilirubin level contains a high conjugated level, then an anatomical obstruction or neonatal hepatitis is the most likely cause. This must always be investigated.

# **CAUSES OF NEONATAL JAUNDICE**

# PHYSIOLOGICAL JAUNDICE

The haem load reaching the liver from the breakdown of RBC is transiently greater than the liver's capacity to conjugate it.

## • HAEMOLYTIC JAUNDICE

- o Maternal-fetal blood group incompatibility (rhesus, ABO, Kell, Duffy etc).
- o Extravascular haemolysis reabsorption of haematoma and petechiae.
- Congenital disorders of the red cell congenital spherocytosis, haemoglobinopathies, glucose 6-phosphate dehydrogenase deficiency, pyruvate kinase deficiency.

#### INFECTIONS

- o Bacterial (generally Gram negative E. coli, Klebsiella, Pseudomonas)
- Viral hepatitis, herpes, cytomegalovirus, rubella, other virus infections
- Other infections toxoplasmosis, syphilis

### OBSTRUCTIVE JAUNDICE

- Congenital atresia of the bile ducts
- Choledochal cyst
- Inspissated bile or cholestatis syndrome
- Cystic fibrosis
- Alpha-1-antitrypsin deficiency

### OTHER CAUSES OF NEONATAL JAUNDICE

- Hypothyroidism
- Galactosaemia
- Breast milk jaundice
- Polycythaemia
- o Drugs sulphonamides, vitamin K and analogues (vitamin K1 excluded) novobiocin
- Hereditary hepatic enzyme deficiencies Crigler-Najjar hyperbilirubinaemia, Gilbert's syndrome, Dubin-Johnsonn syndrome, Rotors syndrome
- Oxytocin in labour

## CLINICAL ASSESSMENT OF THE JAUNDICED INFANT

Jaundice can be readily detected in the newborn when the serum level is 80-90umols/l. A simple and useful method of assessing the degree of jaundice is Kramer's rule (Kramer 1969).

KRAMER'S RULE		AVERAGE SERUM INDIRECT BILIRUBIN
ZONE	JAUNDICE	(PER UMOL)
1.	Limited to head and neck	100
2.	Over upper trunk	150
3.	Over lower trunk and thighs	200
4.	Over arms, legs, below knee	>250

Use this rule to assess if investigations are required. Calculate age of infant in hours, **do not use hospital days but work out age from date and time of birth.** If the "estimated Kramer level" suggests that high levels of hyperbilirubinaemia may develop, if the rate of rise is maintained, then investigations are indicated. The age at which jaundice is first present is important.

## CLINICAL MANAGEMENT OF JAUNDICE (HYPERBILIRUBINAEMIA)

All infants considered to have clinically significant jaundice must be assessed appropriately with thorough physical examination and careful history taking. Jaundice due to specific conditions such as infection, drug interference etc. must be managed according to the diagnosis.

<u>Phototherapy</u> should be used to avoid <u>exchange transfusion</u> in situations where this may become necessary. Infants born at 35-37 weeks gestation have been noted to be at risk for sequelae from severe hyperbilirubinaemia at total serum bilirubin levels lower than those of more mature infants.

The presence of risk factors also need to be taken into account when assessing the serum bilirubin level at which an infant should be given phototherapy and exchange transfusion.

In using the guidelines for phototherapy and <u>exchange transfusion</u> listed, the direct reacting (or conjugated) bilirubin should not be subtracted from the total. In unusual situations where the direct bilirubin level is 50% or more of the total, there are no good data to provide guidance for therapy. Treatment in these cases should be discussed with the consultant neonatologist.

## RISK ASSESSMENT BEFORE DISCHARGE

Assessment of the risk of severe hyperbilirubinaemia should be made on all infants prior to discharge. Infants discharged before 72 hours are likely to still have a rising total serum bilirubin level. The best documented method for assessing the risk of subsequent hyperbilirubinaemia is to measure total serum bilirubin and plot it on a nomogram .

- Infants in the low risk zone are at very low risk of developing severe hyperbilirubinaemia.
- Infants in the high risk zone and those whose total serum bilirubin bilirubin level is crossing centiles upwards, should have their management discussed with the consultant neonatologist or senior registrar.

Follow-up assessment of an infant's level of jaundice after discharge by either a doctor or a visiting midwife would be appropriate when a rise in jaundice levels into the high risk zone is considered possible.

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