



Prolonged neonatal jaundice and its management

Introduction

Jaundice is a common condition in newborn babies and in most cases is harmless. It is generally the result of a build up of unconjugated bilirubin in the blood as part of normal metabolic processes as red blood cells are broken down while liver function is immature. For most babies the level of jaundice is mild and is physiological, and jaundice resolves as liver function develops, generally within two weeks of birth.

However, there are times when neonatal jaundice requires medical attention, and early identification and management of pathological jaundice is crucial for three reasons:

- At high levels, unconjugated bilirubin is neurotoxic in the newborn; it can cause irreparable neurological damage (kernicterus) and death.
- Jaundice may be a symptom of an underlying metabolic or endocrine disease that needs management.
- Prolonged jaundice, or jaundice with dark urine or pale stools, may be a symptom of underlying liver
 disease that needs urgent management. For example, biliary atresia may present with prolonged
 jaundice in the neonatal period. If it is diagnosed promptly and the baby has surgical treatment, the
 outlook is good. Surgery must be performed as soon as possible, and if it is delayed beyond six
 weeks after birth, there may be severe, permanent liver damage, potentially resulting in cirrhosis and
 the need for transplantation.

The Children's Liver Disease Foundation (CLDF) provides resources to support the diagnosis of biliary atresia through its Yellow Alert website.

Causes of neonatal jaundice

There are many causes of jaundice in the newborn and clinicians need to be confident in identifying and managing them:

- Physiological jaundice. This is common: it affects around 60% of babies and 80% of premature babies, typically develops two to three days after birth and resolves without treatment by the time the baby is two weeks old. In around 5% of babies the jaundice is severe enough to need treatment.
- Liver disease. Biliary atresia and other primary liver diseases may cause pathological neonatal jaundice including prolonged jaundice.
- Haemolysis, for example in hereditary spherocytosis, and ABO or rhesus incompatibility.
- Infection. Infection with viruses such as hepatitis B, hepatitis C and cytomegalovirus may cause pathological jaundice in the newborn.
- Congenital hypothyroidism and other metabolic conditions.

Assessment of neonatal jaundice

NICE guidance¹ provides clear advice on the identification and management of jaundice in the neonatal period. All babies should be examined by visual inspection for jaundice at every clinical contact, especially during the first 72 hours after birth.

Jaundice developing within the first 24 hours of birth may be a sign of more serious disease. The serum bilirubin level should be measured urgently (within 2 hours), bilirubin levels monitored, and urgent medical review arranged.

For babies developing suspected or obvious jaundice more than 24 hours after birth, the bilirubin level should be measured urgently (within 6 hours).

For every baby with prolonged jaundice assessment should include consideration of:

- Method of feeding and weight gain (include birth weight and current weight)
- Urine colour/recent wet nappies
- Colour of stool/delayed passage
- Lethargy and sleep/wake/feed behaviour
- Seizures and abnormal movements
- Bleeding/bruising
- Family history
- Blood / liver and metabolic disorders
- Cystic fibrosis
- Antenatal history
- Maternal drug history/infection/USS and blood group

The management of neonatal hyperbilirubinaemia is guided by threshold tables that take into account the bilirubin level and the baby's age and are provided in NICE guidance¹.

Management may include phototherapy or more specialist treatments. Further tests are directed at finding the cause of the jaundice and may include blood packed cell volume, blood group (mother and baby), DAT (Coomb's test), full blood count and blood film, and blood glucose-6-phosphate dehydrogenase levels.

The importance of diagnosing prolonged neonatal jaundice

Prolonged jaundice is defined as jaundice persisting beyond two weeks after birth in a full-term baby and three weeks in a pre-term baby (gestational age less than 37 weeks).

Prolonged jaundice points to a potentially serious metabolic, hepatic or other pathological condition and needs urgent further examination and investigation which may be performed in primary care or through specialist referral depending on the situation.

If a baby has prolonged jaundice, community practitioners must ensure that:

- conjugated bilirubin and a split bilirubin test have been done on an urgent basis, whether this is by
 the hospital team or by the midwife. Conditions causing prolonged jaundice such as biliary atresia
 must be identified and treated as soon as possible to avoid permanent liver damage.
- a history or examination has been made for pale chalky stools and/or dark urine that stains the nappy.

Further investigations may be done in primary care, or may follow specialist referral:

- Take a full blood count and liver function tests
- Take a blood group test (mother and baby) and DAT (Coombs' test)
- Take a urine culture
- Ensure that routine metabolic screening, including screening for congenital hypothyroidism, has been successfully carried out.

Any baby with a conjugated bilirubin level greater than 25 micromol/l may have serious liver disease and should be referred for urgent specialist assessment.

If any jaundiced baby is unwell and/or not progressing normally then the infant should be referred to a paediatrician for investigation.

Why examine urine and stool colour?

Stool and urine colour are valuable pointers to the severity and to the cause of jaundice in a newborn baby:

- Normally a baby's urine is colourless: persistently yellow urine which stains the nappy can be a sign
 of liver disease:
- Normally a baby's stools are green or yellow: persistently pale coloured stools may indicate liver disease.

If the stools and urine in a jaundiced baby are abnormal in colour, the baby should be referred to a paediatrician immediately.

The colour of babies' stools show considerable natural variation. However a helpful stool colour identification chart that can help diagnose abnormally pale stool colour in the neonate can be downloaded from the CLDF website.

Information for parents

There is anecdotal evidence to suggest that when neonatal jaundice is explained antenatally, parents are less anxious if their baby does become jaundiced. In addition parents are more knowledgeable about spotting red flags in jaundiced babies and have a better understanding of when to contact the GP or midwife for advice.

The Children's Liver Disease Foundation produces an information leaflet for parents entitled "Jaundice in the newborn baby" which can be downloaded from the CLDF website

The Children's Liver Disease Foundation has a range of leaflets providing information for parents on liver disease in children. These can be accessed and downloaded from the CLDF information on liver diseases page.

Summary: when to refer babies with neonatal jaundice

The following should be referred to a paediatrician, urgently when needed:

- A baby presenting with jaundice within the first 24 hours after birth.
- A baby with jaundice who is unwell and/or not progressing normally.
- A baby with jaundice with abnormal pale stools and/or urine colour at any age.
- Any baby where the conjugated bilirubin is greater than 20% of the total bilirubin or greater than 25 micromol/l.
- Any baby with prolonged, unexplained jaundice.
- Any baby with jaundice whose bilirubin level is within the NICE treatment threshold for gestational age.

References

1. NICE CG98: Jaundice in newborn babies under 28 days NICE. First published 2010, updated May 2016.