# Management of Neonatal Jaundice in the Paediatric department/Ward Guideline

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| Brief Summary of Document: | Ensuring that all HDUHB Women and Children's Directorate staff utilise the clinical pathway for management of Jaundice. The management and treatment includes the measuring and monitoring of Bilirubin and escalation for treatment. |
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| Scope | This clinical guidance is for use with all babies admitted with jaundice.<br>It is intended to support all employees within the Women and children directorate involved in the care and management of jaundice in conjunction with the NICE guidance for jaundice in new-born babies under 28 days (CG98) |

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| To be read in conjunction with: | Jaundice in new-born babies under 28 days NICE Clinical Guideline October 2016 (CG98) |

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| Owning group | Women and Children's Written Documentation Review Group |

| Reviews and updates | | |
|---|---|---|
| Version no: | Summary of Amendments: | Date Approved: |
| 1 | New Guideline | 14.09.2017 |
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Glossary of terms

| Term | Definition |
|---|---|
| DCT | Direct Coombs test |
| | |
| | |

| Keywords | Neonatal jaundice |
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## Contents

## 1. INTRODUCTION

Approximately 60% of term and 80% of preterm babies develop jaundice in the first week of life, and about 10% of breastfed babies are still jaundiced at 1 month of age. In most babies with jaundice there is no underlying disease, and this early jaundice (termed 'physiological jaundice') is generally harmless. However, there are pathological causes of jaundice in the new-born, which, although rare, need to be detected. Such pathological jaundice may co-exist with physiological jaundice.

Neonatal jaundice refers to yellow colouration of the skin and the sclera (whites of the eyes) of new-born babies that results from accumulation of bilirubin in the skin and mucous membranes. This is associated with a raised level of bilirubin in the circulation, a condition known as hyperbilirubinemia.

## 2. SCOPE

This clinical guidance is for use with all babies admitted to the paediatric department/Ward.
It is intended to support all employees within the Women and children directorate involved in the care and management of prolonged jaundice in conjunction with the NICE guidance for jaundice in new-born babies under 28 days (CG98)

## 3. AIM

The aim of guideline is to ensure best practice in the care management of jaundice, including diagnosis and treatment. It aims to help detect or prevent very high levels of bilirubin, which can be harmful if not treated.

## 4. OBJECTIVES

Ensuring that all HDUHB Women and Children's Directorate staff utilise the clinical pathway for management of Jaundice. The management and treatment includes the measuring and monitoring of Bilirubin and escalation for treatment.

## 5. INTRODUCTION

Bilirubin is a breakdown product of the red cells in the blood. Red cell breakdown produces unconjugated (or 'indirect') bilirubin, which is mostly bound to albumin. Unconjugated bilirubin is metabolised in the liver to produce conjugated (or 'direct') bilirubin, which then passes through the gut and is excreted in the stool. Bilirubin can be reabsorbed again from stools remaining in the gut.

New-born babies' red blood cells have a shorter lifespan than those of adults. The concentration of red blood cells in the circulation is also higher in new-born than it is in adults, so bilirubin levels are higher than they are later in life. The metabolism, circulation and excretion of bilirubin is also slower than in adults. This a degree of hyperbilirubinemia occurring as a result of this normal physiological mechanism is common in new-born babies and usually harmless. It is difficult to tell which babies are at risk of developing high levels of bilirubin that could become dangerous, or who have a serious problem as the explanation for their jaundice, which is why this guideline has been developed.

Physiological jaundice refers to the common, generally harmless, jaundice seen in many newborn babies in the first weeks of life and for which there is no underlying cause.

Beware of Jaundice within first 24 hours of Life or persistent beyond 14 days in term babies or 21 days in preterm babies .Jaundice may also have other, non-physiological, causes, including blood group incompatibility (most commonly Rhesus or ABO incompatibility), other causes of

haemolysis, sepsis, bruising and metabolic disorders. Gilbert syndrome and Crigler–Najjar syndrome are rare causes of neonatal jaundice and are caused by liver enzyme problems. Deficiency of a particular enzyme, glucose-6-phosphate dehydrogenase (G6PD), can cause severe neonatal jaundice. G6PD deficiency is more common in certain ethnic groups and is familial. Congenital obstruction and malformations of the biliary system, such as biliary atresia, cause an obstructive jaundice with conjugated hyperbilirubinemia. This condition needs specialist investigation and early surgical treatment, preferably before 8 weeks of life.

Breastfed babies are more likely than bottle-fed babies to develop physiological jaundice within the first week of life, but the appearance of jaundice is not a reason to stop breastfeeding.

The reasons for the association between breastfeeding and neonatal jaundice have not yet been fully explained but may include inadequate breastfeeding support leading to a reduced intake, sluggish gut action leading to an increase in the entero-hepatic circulation of bilirubin, or unidentified factors in breast milk. Finally, it may be that there is a relative reduction of bilirubin levels in formula-fed babies due to increased clearance of bilirubin from the gut.

### 5.1 BILIRUBIN, ENCEPHALOPATHY AND KERNICTERUS

High levels of unconjugated bilirubin (lipid soluble, and easily crosses blood brain barrier) can cause in the short-term, temperature instability, fits, apnoea, collapse and death. In the long-term, high frequency deafness, choreo-athetoid cerebral palsy and severe learning problems may occur.

This condition is Kernicterus; the name given to the yellow staining of the basal ganglia found at autopsy in bilirubin toxicity. Hopefully it should never be seen!
Intervention aims to prevent these problems.

The level of unconjugated bilirubin which can be considered safe depends on gestational age and also the general condition of the infant. Acidosis and hypoxia displace bound bilirubin from albumin, allowing more bilirubin to cross the blood brain- barrier, thus lowering the threshold for treatment. In the well term infant, problems are rarely seen below about 360 micromols/l. In the pre-term infant the blood brain barrier is less developed, and bilirubin can be pathological at lower levels.

## 6. INVESTIGATIONS

Baseline for all jaundice

- History & full examination, e.g. the spleen is often palpable in haemolysis or sepsis
- FBC and film and reticulocyte count
- Group and Direct Coombs test (DCT) (interpreted in relation to mum's blood group)
- Total bilirubin

**Additional investigations** as suggested by history
More investigations are dictated by suspected cause i.e. Blood cultures and septic screen Urine culture and /or G6PDH screen.

All jaundice admissions should be clerked and admitted on the prolonged jaundice pathway (Appendix 1)

## 7. MANAGEMENT OF JAUNDICE

Phototherapy is the main treatment for unconjugated hyperbilirubinemia.

**The formula**:
Use NICE treatment threshold graphs (see Appendix 2)

**NOTE:**
In cases of Haemolytic Anaemia use a chart that is for babies of 1 week lower gestation.
Phototherapy needs to be started immediately and exchange level is dependent on the rate of rise rather than absolute level as per guideline for
Management of jaundice in first 24 hours – see below

If the infant is sick (HIE, hypoxia, acidosis, hypercapnia, hypoglycaemia, infection) lower thresholds for treatment are necessary therefore use a chart of 1 week lower gestation e.g. for a baby 0f 30 weeks gestation use a chart for 29 week GA.

Phototherapy converts bilirubin to a soluble form allowing its renal excretion
**Remember, to ensure you have the correct chart for the baby's gestation.**

### 7.1 Indications for phototherapy

Phototherapy should be started in any baby whose serum or transcutaneous bilirubin plots above the treatment line on the NICE jaundice treatment chart (available on SharePoint and via Google) or if the TCB is >250 or within 50micromol/L of the treatment line.

### 7.2 How to Start Phototherapy

**If serum bilirubin is above the treatment line but more than 50 micromol/L below the threshold for exchange transfusion start single phototherapy.**
Short breaks of up to 30 minutes for breast feeding can be allowed. Use clinical judgement.
Repeat serum bilirubin* 4-6 hours after starting phototherapy. Then every 6-12 hours if level stable or falling. Switch to continuous multiple phototherapy if level is not stable or falling. Monitor hydration by checking for wet nappies.
**If serum bilirubin less than 50 micromol/L below the threshold for exchange transfusion or if haemolytic disease of the new-born is suspected:**

- **Start continuous multiple phototherapy**

### 7.3 Types of Phototherapy

- **Overhead lights**

These are generally the first choice
Additional overhead lights can be added to increase the level of phototherapy delivered
Babies can get cold under overhead lights and may need to be in an incubator to maintain their temperature.

- **Biliblanket**

This is a mattress which delivers phototherapy and allows the baby to be covered with a blanket whilst undergoing treatment.
Only delivers single phototherapy but can be used in conjunction with overhead lights to deliver multiple phototherapy.

### 7.4 Caring for a baby receiving phototherapy

- Ensure as much skin exposure as possible. Lay on an open nappy. Remove hat.
- Ensure the eyes are covered with appropriate phototherapy eye protection.
- If using overhead lights, the baby will probably need an incubator or overhead heater to keep warm. The temperature should be measured 4-hourly.

Phototherapy itself does not increase fluid requirements; however dehydrated babies often have more severe jaundice and an assessment of feeding and hydration should be made. If the SBR is particularly high or is not reducing despite phototherapy send U+Es and consider NG top ups

### 7.5 Managing a baby requiring phototherapy

- The baby should have as much skin exposed as possible, so lay the baby on an open nappy with no hat (unless ventilated).
- The baby will probably need an incubator or if using bili-blanket overhead heater to keep warm.
- The temperature should be measured 4-hourly.
- Explain fully to the mother before starting.
- The mother should be allowed to handle the baby for feeding, changing and cuddling the baby out of the phototherapy with eyes uncovered, as long as there is no immediate risk of having to do an exchange transfusion. However, baby should not be out of the phototherapy for prolonged periods.
- Phototherapy itself does not increase fluid requirements; however dehydrated babies often have more severe jaundice and an assessment of hydration should be made. If the SBR is particularly high, then electrolytes should be checked and if dehydration is suspected, NG feeds or IVI considered.
- Repeat serum bilirubin measurement 4-6 hours after initiating phototherapy
- Repeat serum bilirubin measurement every 6- 12 hours when the serum bilirubin level is stable or failing ( as per NICE guidance )
- For Nursing care management please follow the nursing care plans.

Consider intense phototherapy (often known as double phototherapy) if –

- the serum bilirubin level is rising rapidly (more than 8.5 micromol / litre per hour)

- the serum bilirubin is at a level within 50 micromol / litre below the threshold for which exchange transfusion is indicated after 72 hours or more since birth (see threshold table and the treatment threshold graphs)

- the bilirubin level fails to respond to initial phototherapy (that is, the level of serum bilirubin continues to rise, or does not fall, within 6 hours of starting phototherapy).

- Intense phototherapy is defined as - Phototherapy that is given with an increased level of irradiance with an appropriate spectrum. Phototherapy can be intensified by adding another light source or increasing the irradiance of the initial light source used.(NICE 2016)

### 7.6 Parent information

Offer parents or carers information about neonatal jaundice. This information should be provided through verbal discussion backed up by written information.

### 7.7 Stopping Phototherapy:

- Stop phototherapy once serum bilirubin has fallen to a level at least 50 micromol / litre below the phototherapy threshold.

- Recheck SBR (for rebound) within 24 hours after stopping phototherapy, except in sick or premature babies, when a level should be repeated earlier – 12 hours after stopping phototherapy. Babies do not necessarily have to remain in hospital for this to be done if well enough to go home.

## 8. PROLONGED JAUNDICE

Prolonged jaundice, that is jaundice persisting beyond the first 14 days or more than 21 days in preterm, is also seen more commonly in term breastfed babies. Prolonged jaundice can be a clue to serious underlying liver disease and should be assessed carefully.

### 8.1 Causes of prolonged jaundice:

- Persistence of unconjugated jaundice from early neonatal period:
  - Haemolytic jaundice (of any aetiology)
  - Infection, including UTI
- Rare causes of unconjugated jaundice:
  - Inborn errors of metabolism (Galactosaemia, tyrosinemia, lipid-storage disorders, and others)
  - Hypothyroidism
  - Drugs
  - Crigler-Najjar
  - Gilbert's
  - Intestinal obstruction
  - Cystic fibrosis
- Conjugated jaundice
- Breast milk jaundice

### 8.2 Investigations of Prolonged Jaundice

When accessing the baby for underlying disease, consider whether the following tests are clinically indicated. In Acute paediatrics please use the pathway for management of prolonged Jaundice (Appendix1)

- History & examination to elicit cause, please check the colour of stool from the referring midwife or health visitor and request to bring a stool nappy with the family)
- Base line investigations:
- Split SBR (total and direct SBR) - the direct SBR is the conjugated fraction - a value of >20% of the total or of >25 mmol/l is suggestive of an obstructive or hepatic cause for the jaundice.
- FBC. Group and Coomb's. Blood film.
- LFT & coagulation screen
- TFTs (If the Heel Prick Neonatal Screen result cannot be obtained by ringing the UHW Neonatal Screening Lab)
- Urine culture
- Further septic screen, if unwell
- Urine for reducing substances

### 8.3 Prolonged Conjugated jaundice:

This is usually picked up because of prolonged jaundice with a high direct fraction. There may be a history of pale stools and dark urine, which should ring alarm bells for biliary atresia and other causes of obstructive jaundice.

### 8.4 Further investigations in case of conjugated jaundice:

- Hepatitis A, B and C
- Gal-1-PUT
- α1-antitrypsin phenotype. (NB, since α1 antitrypsin is an acute phase reactant, it may be raised in any intercurrent illness and may be normal even if there is a deficiency. Hence the need for the typing.)

- TORCH screen
- Serum amino acids and urine organic acids, and urine organic acids
- X-ray spine (hemivertebra in Alagille syndrome)
- Liver USS (to exclude choledochal cyst, and look for signs of biliary atresia)
- Radio-isotope scan of the liver to detect biliary atresia
- Eye examination (posterior embryotoxon)

## 9. BREAST MILK JAUNDICE

This is a diagnosis of exclusion. Excessive investigations may cause anxiety to parents, and lead mother to stop breast feeding. Jaundice is not a reason to stop breastfeeding but fluids may need to be increased.

If the baby is well and is normal on examination, and has normally pigmented stools, check the baseline investigations (see above). If these are normal, no further investigations are needed unless the clinical picture changes or the jaundice clinically becomes darker, in which case, a further split SBR should be checked. If the jaundice does not resolve in a week, then the split SBR should again be repeated, but if it remains unconjugated and the baby is well, no further tests are needed

## 10. IMPLEMENTATION AND TRAINING

All Healthcare Professionals involved in care of jaundiced babies must be appropriately trained in all aspects of education on use of the treatment threshold graphs and baseline assessment and nursing care. Please use presentation slide from NICE tools and resources 15 (appendix 3)

## 11. REFERENCES

Jaundice in new-born babies under 28 days NICE Clinical Guideline (CG98) updated 26 October 2016

## 12. Appendix 1 - Prolonged Jaundice Pathway

**PATIENT IDENTIFICATION LABEL:**
Name
Permanent Address & Postcode
NHS Number
Hospital Number
Date of Birth

**Women and Children's Directorate**
**Prolonged Neonatal Jaundice Management**

GIG CYMRU NHS WALES
Bwrdd Iechyd Prifysgol Hywel Dda University Health Board

**Prolonged jaundice**

If term > 14 days
If premature > 21 days

(Premature is < 37/40 at birth)

→

**Clinical assessment**
- Unwell
- Failure to Thrive
- Pale Stools (staff/paediatrician to visualise stool colour)
- Dark Urine
- Increasing Jaundice
- Bleeding / Bruising

If any of above present then admit immediately and consider conjugated hyperbilirubinaemia pathway

**First Line Investigations**

- FBC
- Total and Split Bilirubin
- Check newborn screening performed, if not TFTs

→ **If conjugated bilirubin is > 25 µmol/L**

Then follow conjugated hyperbilirubinaemia pathway

→ **Admit**
**Consider arranging USS Abdomen**
**Inform on-call consultant**

→ **Investigate for unconjugated hyperbilirubinaemia**
**Check treatment threshold graphs**

→ **If conjugated bilirubin is < 25 µmol/L**

And other investigations are normal

→ **If total bilirubin > 200 µmol/L**

**Any sick infant and those with pale stools should be discussed early with Liver unit, Birmingham Children's Hospital**

**NO**

**YES**

**If unconjugated bilirubin < 200 µmol/L**
Exclusively breast fed baby thriving well

**No further follow up unless clinically indicated**

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**PATIENT IDENTIFICATION LABEL:**
Name
Permanent Address & Postcode
NHS Number
Hospital Number
Date of Birth

## Conjugated Hyperbilirubinaemia Management

These guidelines are only applicable to infants with conjugated hyperbilirubinaemia.
**Early discussion with Liver Team in Birmingham Children's Hospital essential (don't necessarily wait for the results of all investigations prior to discussion).**

**First Line Investigations**

Pre-feed BM within first 24 hours of admission

FBC and Reticulocyte Count
Blood Group
Coomb's
INR
Prothrombin Time
APTT
Fibrinogen
Urea and Electrolytes
Bicarbonate
Calcium
Phosphate
Total Bilirubin
Conjugated Bilirubin
LFT's
AST
Gamma GT
Lipid Profile

If INR is prolonged give 300 micrograms/kg Vitamin K IV and repeat INR at 1-2 hours. If still prolonged contact a Liver Unit.

→ **Infections**
Blood Cultures
Urine Dipstick and culture
Urine CMV
Serology (IgM to Toxoplasma, Rubella, CMV and Herpes)
Hepatitis A, B and C Serology

→ **Metabolic**
Immunoreactive Trypsin (up to 8 weeks) or sweat test
Galactose-1-Phosphate Uridyl Transferase
α1 Antitrypsin Level and Phenotype
Plasma and Urine Amino acids
Urine Organic Acids (Succinyl Acetone)

→ **Endocrine**
TFT's
Cortisol (if low consider a short synacthen test)

→ **USS Abdomen**

**After a 4 hour fast, see if gallbladder present and if there is a choledochal cyst**

**Observe stool sample and save for consultant review**

→

**Second Line Investigations**
**(After discussion with Liver Team)**

Hepatobiliary Scintigraphy

Pre-treat with phenopharbitone 5mg/kg nocte for at least 3/7 and continue until 24 hours post-isotope if there has been no excretion before then

Liver biopsy
Syphilis Serology
Viral PCR (Herpes, CMV)
Eye examination for embryotoxon, chorioretinitis and septooptic dysplasia
XR Spine for butterfly vertebrae
Cardiology opinion if mumur heard

Tests for rare disorders. Lactate, ammonia, pyruvate. Very long chain fatty acids. Urine and serum for inborn errors of bile salt metabolism. Acyl carnitines. α Fetoprotein. Isolectric focusing for transferring. White cell enzymes either glycogen or lysosomal storage. CSF for protein and lactate. Tubular reabsorption of phosphate. Ferritin and transferring saturation. MRI Head. Muscle biopsy for mitochondria cytopathy. Bone marrow for storage disorders. Skin biopsy for fibroblast culture.

| | | | | | |
|---|---|---|---|---|---|
| **NURSING ENTRY:** Weight:- ( centile) | | | Head Circumference (centile) | | |
| **OBSERVATIONS :** | **TEMP** | **HEART RATE** | **RESP RATE** | **O2 SATS** | **CRT** |
| **History** | | | | | |
| Gestation | | | Day of life | | |
| Colour of stools | | | Colour of urine | | |
| Feeding (breast/bottle/mixed) | | | weight gain/loss | | |
| Family history/ethnicity | | | Guthrie(Heel Prick) | | |
| Intercurrent illness | | | Others (Maternal Concerns) | | |
| **General Examination** | | | | | |
| Dysmorphic features | | | Looks well/unwell | | |
| Jaundice | | | palor | | |
| **Abdomen examination** | | | | | |
| Liver: | | Spleen: | | Ascites: | |
| Other examination: Skin:<br>Eye: Cataract | | | | | |

**INVESTIGATIONS TO BE COMPLETED:-** **Total and Conjugated bilirubin (one green top)*** **FBC (one purple top)****

| Date | Hb, neutrophils | SBR | Conjugated Bilirubin | Urine M,c&S |
|---|---|---|---|---|
| | | | | |

**Repeat investigations:-**

| Date | Hb, neutrophils | SBR | Conjugated Bilirubin | Urine M,c&S |
|---|---|---|---|---|
| | | | | |

Document Communication with parents:-

Final action:-

1. Investigations normal—Discharged
2. Repeated investigations—Normal—Discharged
3. Abnormal investigations—Referred to Liver Unit, Birmingham Children' Hospital

Abnormal results/investigations --- further investigate as per the guideline

| COMPLETED BY | Print Name | signature | Date | Time |
|---|---|---|---|---|
| | | | | |

**Abnormal result**

*conjugated bilirubin >20 micromol/l (if total <85 micrommol/l) or Conjugated bilirubin >20% total (if total >85 micrmmol/l)or total bilirubin >200 )

**Hb<10, Neutrophils<1 Urine MC&S shows pure growth

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## First Line Investigation Sheet: (Only for Conjugated Hyperbilirubinemia)

| | Test | Date taken | Result |
|---|---|---|---|
| **Haematology** | FBC | | HB: WBC: PLT: RETICS: |
| | Coagulation | | INR: PT: Aptt: Fib: |
| | Group & Coombs | | Gp: Coombs: |
| **Biochemistry** | U/E | | Na: K: Urea: Cr: |
| | Bilirubin | | Total: Conj: Unconj: |
| | LFT | | ALT: GGT: ALP: |
| | | | Total Protien: Albumin |
| | Cholesterol/Trig | | Chol: Triglyceride: |
| | Bone profile | | Ca: Phosphate: |
| | Blood Glucose | | |
| **Microbiology** | Blood Culture | | |
| | Urine Culture | | Microscopy: Culture: Sensitivity: |
| **Virology** | IgM TORCH | | Toxo: Rub: CMV: Herpes: |
| | Hepatitis A B C | | Hep A: Hep B: Hep C: |
| | Urine CMV | | |
| **Metabolic** | Sweat Test/IRT | | |
| | Gal 1 put | | |
| | Alpha 1 antitrypsin | | Level: Phenotype: |
| | Plasma amino acids | | |
| | Urine amino acids | | |
| | Urine organic acids | | |
| | Ward test Urine | | |
| **Endocrine** | Thyroid function test | | T3 : T4: TSH: |
| | Cortisol (4 hrs after fast) | | |
| | Short Synacthen Test* | | |
| **Radiology** | Abdominal USS (fasting) | | |
| | CXR/Spine Xray* | | |
| | Radioisotope scan* | | |
| **Histology** | Liver Biopsy* | | |
| **others** | Ophthalmology review | | |
| | Stool Colour | | |

***Second line investigation**

**Discussion with Birmingham Liver Unit:** **Liver.Direct@bch.nhs.uk**
**Tel: 0121 333 9999 / Bleep 55200**
**Fax: 0121 333 8251**

**Outcome of discussion:**

| | Print Name | Signature : | Date | Time |
|---|---|---|---|---|
| COMPLETED BY | | | | |

**References:**


1. NICE guideline – neonatal jaundice CG98 updated in 2010
2. BSPGHAN revised guideline for Conjugated Hyperbilirubinemia Feb 2012
3. Prolonged Jaundice Guideline – Nottingham Children's Hospital 2013


Developed by : Dr Sabyasachi Chowdhry, Paediatric registrar, Bronglais General Hospital, Hywel Dda University Health Board 2016 updated 2019
Approved By : W&C Policy group June 2019

## 13. Appendix 2 - Treatment Threshold Graphs

Copy of
treatment-threshold-g

## 14. Appendix 3 - Training Slides (NICE CG 98)

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