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# Learning from claims: hyperbilirubinaemia and kernicterus

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## ABSTRACT

We examined claims made against the National Health Service (NHS) involving neonatal jaundice in order to determine whether there were lessons that could be learnt from common themes.

This was a retrospective anonymised study using information from the NHS Resolution database for 2001–2011.

Twenty cases (16 males) had sufficient information for analysis. Fifteen had confirmed cerebral palsy and two young children had damage to the globus pallidus without confirmed CP. In three cases, the outcome was uncertain. Two were extremely preterm, five were born at 34–36 weeks' gestation. Jaundice was typically present very early in life; in four cases, it was noted at less than 24 hours of age, and in 14 cases, it was first noted on the second to third day. There was a lag between recognition and readmission, with a range of 26–102 hours. The peak serum bilirubin level was over 600 µmol/L in all the babies born at term. An underlying diagnosis was found in all but two; six had glucose-6-phosphatase deficiency (one also had Gilbert's syndrome); five were diagnosed with ABO incompatibility; three with Rh haemolytic disease; one with spherocytosis and three preterm. The total cost of these claims by August 2017 was almost £150.5 million. This figure is likely to rise.

These data show that, in the group who litigate, babies who develop kernicterus generally have an underlying diagnosis. We recommend adherence to the National Institute for Health and Care Excellence guideline that recommends measuring the bilirubin level within 6 hours in all babies who are visibly jaundiced.

## INTRODUCTION

The management of neonatal jaundice is targeted at preventing kernicterus, which can result from high levels of unconjugated bilirubin. Kernicterus is a lifelong and serious disability. Individuals with kernicterus typically have dyskinetic tetraplegic cerebral palsy and bilateral sensorineural deafness, often with an upgaze palsy.

The lifetime costs of care for people with kernicterus are considerable and are reflected in the value of successful claims. Prior to March 2017, these were on average £12 million per case, but settlements are now significantly more due to the change in the personal injury discount rate.

Neonatal jaundice is very common, and most babies have a transient and harmless rise in bilirubin levels due to a relatively high haemoglobin load, immature liver conjugation systems and temporary recirculation via the enterohepatic route while feeds are being established. Considerable time,

## What is already known on this topic?

- ▶ Kernicterus is the cause of life-long disability due to high levels of unconjugated bilirubin; the exact level of bilirubin, which is neurotoxic, is not known, nor is the duration of exposure.
- ▶ Neonatal jaundice is extremely common, and using current National Institute for Health and Care Excellence guidelines, around 5%–10% of babies born at term are treated with phototherapy.
- ▶ Extreme hyperbilirubinaemia (levels above 510 µmol/L) is rare, occurring in 7 babies per 100 000 births in the UK.

## What this study adds?

- ▶ The cost of handling 20 claims made against the National Health Service between 2001 and 2011 was £150 million, and this figure is likely to rise.
- ▶ Half of the children with kernicterus were not of Caucasian ethnicity; six had glucose-6-phosphatase deficiency, five had ABO incompatibility and three had Rh disease.
- ▶ The peak serum bilirubin was over 600 µmol/L in all the term-born babies with kernicterus; there was a lag between recognition of jaundice and admission of 26–102 hours.

effort and National Health Service (NHS) resource is spent in recognising and managing neonatal jaundice. A National Institute for Health and Care Excellence (NICE) guideline was published in 2010, with recommendations regarding when to measure bilirubin and with threshold levels for intervention. Jaundice is the most common reason for neonatal readmission from home.<sup>1</sup> The NICE jaundice guideline has a lower threshold level for initiating phototherapy at 37 weeks compared with 38–42 weeks,<sup>2</sup> which was probably one of the main contributing factors behind the high readmission rate for babies born at 37 weeks seen in the Battersby study. Whether kernicterus develops at lower bilirubin levels in babies of 37 weeks' gestation compared with those of 38 weeks' gestation or more is not known. Adjusting the thresholds at 37 weeks to match those at 38–42 weeks would reduce the admission rate, reduce separation of mother and baby and may not have any impact on the kernicterus rate.



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Current estimates suggest that around 6% of babies born at more than 35 weeks' gestation are treated with phototherapy<sup>3</sup>; this increases to around 9% at term when screening procedures are used.<sup>4</sup> Extreme hyperbilirubinaemia is rare, with 7 babies in 100 000 being reported to have a bilirubin higher than 510  $\mu\text{mol/L}$  in the UK in 2003–2004.<sup>5</sup> One hundred and twenty (15.2 per 100 000 births) babies were treated with an exchange transfusion in the UK between October 2014 and October 2015, 109 for hyperbilirubinaemia (Gottstein *et al*, BPSU survey, paper in preparation). From these data, a broad estimate of the number of babies who are treated with phototherapy to prevent one case of kernicterus suggests that perhaps as many as 5000 babies are treated to prevent one case. Most babies who are treated with phototherapy in hospital require a short course of treatment and do not ever have bilirubin levels above 350  $\mu\text{mol/L}$ . This extremely high number 'needed to treat' prompted us to examine the NHS Resolution claims management system for hyperbilirubinaemia claims in order to determine whether we could establish any common themes that might inform future management of neonatal jaundice and to establish whether the NICE guidelines had been followed in the cohort born after May 2010.

## METHODS

Research ethics committee approval was not required, as this was a retrospective anonymised study using routinely collected data from the NHS Resolution database. Claims notified to NHS Resolution between 2001 and 2011 alleging that kernicterus was preventable were identified from the NHS Resolution claims management system. Anonymised documentation relating to the claims was reviewed by a consultant neonatologist (JMR) and the NHS Resolution Safety and Learning Lead (Obstetrics) (JB). Information was extracted from letters of claim, letters of response and expert reports. The database was searched for keywords neonatal, jaundice and kernicterus and manually checked by JMR and JB. Information was included from open and closed claims. NHS Resolution collects information only from cases in England.

The following data were extracted where available from the documentation held on the database for each case:

- ▶ Date and time of birth and gender.
- ▶ Gestational age and birth weight.
- ▶ Ethnicity.
- ▶ Maternal and infant blood group, direct antibody test result.
- ▶ Family history of jaundice.
- ▶ Time and date of first discharge.
- ▶ Date and time of recognition of jaundice.
- ▶ Date and time of readmission.
- ▶ Date, time and value of peak serum bilirubin level.
- ▶ Use of sunlight.
- ▶ Date and time of commencement of phototherapy.
- ▶ Date and time of any exchange transfusion.
- ▶ Nature of underlying cause.
- ▶ Presence of clinical encephalopathy.
- ▶ MRI and result.
- ▶ Nature of disability: deafness and/or cerebral palsy.

JMR attempted to identify any specific factors that may have led to the development of kernicterus, in particular, any avoidable factors. Specific comments from staff or parents were noted.

## RESULTS

Twenty-six cases were identified, of these 20 had enough available information for analysis. Seven babies were born after the

introduction of the NICE guidelines in 2010; all were born after the year 2000.

MRI had been done in 15 cases, with confirmation of globus pallidus damage in 13 cases. Fifteen babies were reported to have cerebral palsy, while 12 were reported to have deafness. In five the final neurological outcome was not available, but two of these had globus pallidus damage so are likely to develop cerebral palsy. Information about albumin levels was not generally available.

Sixteen of the 20 cases were male; 10 were of Caucasian ethnicity, 4 Asian, 4 Afro-Caribbean, 1 baby was Greek and in one the ethnicity was not stated. Two babies were extremely preterm at 25 weeks; of the remainder, five babies were born at 34–36 weeks of gestation, 12 at term and in 1 the gestation was not stated. The mature babies were all above 2.5 kg birth weight.

One baby, with probable haemolytic disease and a positive direct antibody test on cord blood, was discharged and not readmitted in the neonatal period. His bilirubin level was not measured in the neonatal period, and he received no treatment. The diagnosis of probable kernicterus was reached with hindsight and accepted; his mother had high levels of anti-D and anti-C antibodies. He had dyskinetic cerebral palsy, high frequency hearing loss and abnormalities in the globi pallidi on MRI of his brain. Very extensive investigations for other causes of dyskinetic cerebral palsy were negative.

Jaundice was recognised to be present early in the life of most of the babies; in four babies, jaundice was reported within 24 hours, and in 14 babies, it was noted on the second to third day. There was typically a lag between recognition and readmission; three babies were not discharged from the neonatal unit (two born at 25 weeks' gestation and one with Rh disease) and one (see above) was never readmitted. In the remaining 15 babies in whom the time of readmission was known, the range of time in hours between recognition and readmission was 26–102 hours, with nine babies being readmitted more than 48 hours after the initial recognition of jaundice. Information about the time of readmission was not available for one baby. Information about transcutaneous bilirubinometry was not available.

Peak serum bilirubin levels were strikingly high in this group. Of the babies between 34–36 weeks of gestation, the peak levels that were reported were 579, 602, 330 and 490  $\mu\text{mol/L}$ . In the term group, the peak levels were all above 600  $\mu\text{mol/L}$  with one baby having a level of over 1000  $\mu\text{mol/L}$ . An underlying diagnosis was found in all but two cases; six babies had glucose-6-phosphatase deficiency (one also had Gilbert's syndrome); five were diagnosed with ABO incompatibility; three with Rh haemolytic disease; 1 with spherocytosis; and 3 with prematurity.

Of the remaining babies, all 19 were treated with phototherapy and 13 had an exchange transfusion. Sunlight was used in eight babies, three of whom were born after the recommendation in the NICE guideline (May 2010) that sunlight should not be used. Mothers frequently reported comments such as 'the midwife said the jaundice was nothing to worry about, and that putting the baby in the window would be sufficient', or 'the midwife advised daylight and regular feeds'.

There were common themes that can be grouped into both clinical and systems failures. The most common clinical failure was a delay in measuring bilirubin in babies with visible jaundice, three of whom had a family history of neonatal jaundice and two of whom were jaundiced from very early in life. Lessons learnt from the past regarding the management of Rh disease seem to have been lost, and in three cases there was a lack of recognition of the importance of a diagnosis of Rh incompatibility antenatally with rising antibody levels and a positive direct

antibody (Coombs) test on cord blood. There was an inappropriate reliance on phototherapy in some cases of Rh disease, with a seeming reluctance to proceed to exchange transfusion in spite of bilirubin levels that were above the threshold. There were system failures, with delays in treatment or admission due to lack of neonatal intensive care unit cots, which were considered serious in at least two cases. Babies were held in inappropriate areas such as Accident and Emergency departments while waiting for a bed, without phototherapy being initiated. Others were transferred from one hospital to another due to lack of beds, or because of a possible need for an exchange transfusion, without phototherapy being initiated.

The cost to the NHS of handling and settling these claims was considerable, with the cost of these 20 cases (to August 2017) currently running at £150.5 million. Claimant solicitor's costs were approximately £6.5 million with £2.2 million defence costs. Future claims are envisaged for medical expenses, travel costs, assistive technology, loss of earnings, education, aids/appliances, physiotherapy and holidays. The current outstanding reserves for these claims is £55 242 500.

## DISCUSSION

This retrospective survey shows that kernicterus is still occurring in England, with approximately two cases a year coming to litigation. Our findings are likely to provide useful messages for similar developed healthcare systems, in which similar failings have been recognised.<sup>6</sup> This is likely to be an underestimate. Seven babies were born during 2010–2011; NICE guidance was published in May 2010. Guidance takes some time to implement, and it is too soon to draw any conclusions regarding the effectiveness of the new guidance. There are data that suggest that the incidence of bilirubin encephalopathy has fallen between 2012 and 2015 (from 1.2 to 0.6 per 100 000 births).<sup>7</sup>

The costs of these claims is likely to rise. This is due to incidents that have already occurred but yet to be reported as a claim, and the outstanding reserve costs for the non-settled claims, which were £55 242 500 (inclusive of damages claimant and defence costs) at the time of the data analysis. A further confounder is the recent change to the personal injury discount rate. Where compensation is paid to claimants for needs that will arise in the future, it is generally accepted that a full payment made at the time of settlement could be invested and result in an overpayment/underpayment over time. As such an actuarial discount factor is applied to account for this. Prior to March 2017, a discount factor of 2.5% was applied. Due to the economic climate of the last few years, this has been changed to –0.75%. As a result, the amount paid out in respect of future losses has increased significantly.

The challenge for maternity services continues to lie in the identification of the rare baby whose bilirubin level is rising rapidly to levels that are potentially neurotoxic, while avoiding overmedicalisation and unnecessary hospital admission in the vast majority. The risk of kernicterus is low, probably around 1 per 100 000 births, whereas jaundice is visible in approximately

60% of all babies born at term. The number of babies admitted to hospital and treated with phototherapy is considerable, often causing separation of mother and baby and incurring substantial costs to the NHS, yet cases of kernicterus are still occurring. These data show that, at least in the group of babies whose parents litigate on their behalf, most of those who develop kernicterus have an underlying medical diagnosis (ie, the jaundice is not physiological) and have bilirubin levels of over 600 µmol/L at presentation at term. There is clearly a balance to be struck between the desire to prevent kernicterus occurring and the desire to avoid unnecessary hospital admission and treatment for mild to moderate hyperbilirubinaemia, which is never likely to prove dangerous. From these data, we suggest that several messages emerge, and these include the importance of ethnicity, family history, continued inappropriate use of sunlight and a delay in measuring bilirubin levels in babies who are recognised to have jaundice. Our analysis was limited because data was extracted from unstructured expert reports, not the original notes, and on occasion important detail was lacking. We agree with Bhutani and colleagues, who urge an 'aviation style' approach to the problem and call for national registries of kernicterus cases.

**Contributors** JMR extracted and analysed the data and wrote the paper. JB extracted the data, designed the spreadsheet and contributed costings and comments to the paper. MU contributed regarding the NHS perspective and to the writing of the paper.

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