

# Kernicterus in Two Generations: A Need for More Aggressive Preventive Measures

Kuti Bankole Peter<sup>1\*</sup>, Ogundele Tolulope<sup>2</sup>, Adeniyi Adewuyi Temidayo<sup>3</sup>, Kuti Demilade Kehinde<sup>4</sup>

<sup>1</sup> Department of Paediatrics and Child Health, Obafemi Awolowo University/Wesley Guild Hospital Unit of the Obafemi Awolowo University Teaching Hospitals Complex, Ile-Ife, Osun State, Nigeria

<sup>2</sup> Department of Paediatrics Wesley Guild Hospital, Ilesa, Nigeria

<sup>3</sup> Department of Paediatrics Wesley Guild Hospital, Ilesa, Nigeria

<sup>4</sup> Department of Paediatrics, Wesley Guild Hospital, Ilesa, Nigeria

## ABSTRACT

Bilirubin induced brain damage is still a common cause of morbidity and mortality in Nigerian infants. No much success has been recorded in decades despite the fact that brain damage from bilirubin toxicity in the newborn is easily preventable with affordable and effective means.

We report a case of a newborn with acute bilirubin encephalopathy. This case was considered of interest because the mother of the baby herself had kernicterus as a newborn now manifesting with features of choreoathetoid cerebral palsy. The baby was brought to our facility late just as how the mother was presented late to a health facility 30 years ago. The unfortunate occurrence of brain damage from bilirubin toxicity in two successive generations calls for urgent attention.

We advocate more aggressive and all-embracing preventive measures including routine home visit of newborns, awareness creation and health education and involvement of all stakeholders to nip this problem in the bud.

**Keywords:** Kernicterus, Neonatal jaundice, Preventive measures

## Introduction

Jaundice occurs in 60-80% of newborn and it's a cause of neonatal morbidity and mortality (1). Kernicterus which result from untreated severe jaundice is described as pathologic staining of the brain by unconjugated lipophilic bilirubin and is one of the most common causes of preventable brain damage in developing countries (1-2). It greatly impairs the quality of life of children and their care-givers, and impedes their ability to fulfil their potentials (3). Kernicterus constitutes a great burden to the health facility and consequently leads to tremendous loss to the nation (3). The most unfortunate aspect of the problem is that though Kernicterus is easily preventable with affordable, efficient and evidence-based means (4), little success has been achieved in this respect in Nigeria (5).

We report the unfortunate occurrence of kernicterus in two successive generations of a mother and her baby who presented at the Wesley Guild Hospital Ilesa (WGH) - a unit of the Obafemi

Awolowo University Teaching Hospitals Complex (OAUTHC), Ile-Ife, Nigeria.

## Case report

Baby OB (male) was admitted to the newborn unit of the WGH, Ilesa, at age 5 days with complaints of Jaundice noticed in the second day of life; poor sucking, activity and cry, and abnormal body movement noticed about 9 hours before presentation. Since the jaundice was noticed, baby had been on oral antibiotics bought over the counter and locally prepared herbal concoction including extract from pawpaw fruits (*Carica papaya*). There is denial of the use of icterogenic agents.

Baby OB was delivered via spontaneous vaginal delivery at home to a 30 year old woman at EGA 34 weeks and mother had pre-labour rupture of foetal membrane 48 hours before delivery. No history to suggest birth asphyxia. Birth weight was however not known.

\* Corresponding author: Kuti Bankole Peter, Department of Paediatrics and Child Health, Obafemi Awolowo University/Wesley Guild Hospital Unit of the Obafemi Awolowo University Teaching Hospitals Complex, Ile-Ife, Osun State, Nigeria. Email: kutitherapy@yahoo.com

The remarkable aspect of this case is that the mother of the baby is a 30 year old housewife who had features of dyskinetic cerebral palsy – choreo-athetoid movement, enamel dysplasia, sensorineural deafness and defective upward gaze. She is handicapped by the condition and has to be supported in the care of the baby as well as her personal grooming by the grandmother. The father is 33 year old artisan (Barber).

Further enquiries about event surrounding the birth of the mother from the maternal grandmother revealed that she (the mother of the baby) was also delivered at home, developed jaundice within 24 of delivery which was managed with local herbal remedies including extract of *Carica papaya* fruit and ampiclox syrup until she was noticed not to be sucking well with poor activities and abnormal limb movement necessitating presentation at a private hospital where she was managed for 5 days before discharge.

At admission, baby OB had features of preterm (EGA 34 weeks) weighed 1.8 kg, deeply icteric, not pale, hypothermic (temperature 36.3°C), had cyclic movement of the limbs, depressed primitive reflexes, sun-setting eyes and poor cry. The other vital signs and systemic examinations were normal.

Investigation done at admission for baby OB revealed packed cell volume 60%, baby's blood group O rhesus+, mother's blood group A rhesus +, total serum bilirubin (tsb) 585µmol/l (34.4mg/dl), conjugated fraction was 85µmol/l ( ), Glucose-6-phosphate dehydrogenase (G6PD) assay was deficient, Direct Coomb's test was negative, Full blood count and electrolytes estimations were normal. Blood culture yielded no growth of any organism.

Assessment of LBW preterm baby with severe neonatal jaundice and features of bilirubin encephalopathy secondary to G6PD deficiency was made. The baby had double volume exchanged blood transfusion, phototherapy and supportive treatment. Parents and other caregivers were counselled to avoid use of icterogenic agents and about the care of the baby including expectations vis-a-vis the bilirubin encephalopathy. The baby was discharged after a week with tsb 68µmol/l to be followed up in the clinic.

## Discussion

Neonatal jaundice (NNJ) is a significant cause of newborn morbidity and mortality reported to contribute to 23% of neonatal deaths from a

tertiary centre in Nigeria (6). Severe neonatal jaundice may cause kernicterus which can result in death and or irreversible brain damage (1-2). Jaundice may also be a manifestation of underlying, potentially life-threatening conditions like sepsis, hypothyroidism and G-6-PD deficiency (1). Jaundice in the newborn should therefore be always treated as neonatal emergency.

There have been many reports on the need for early recognition and appropriate management of newborn jaundice (5, 7-8). Sadly, Kernicterus is still a common cause of death and morbidity in the newborn units of many hospitals in developing countries. Owa *et al* (9) about 30 years ago reported that 9.8% of babies who presented with jaundice at the WGH had overt features of Kernicterus at presentation. More recent observation in the same centre revealed that brain damage from severe hyperbilirubinaemia is still wreaking havoc to our babies. Owa *et al* (5) in a more recent study from our centre reported that 30% of the babies with severe jaundice who had EBT already had features of Kernicterus at presentation as seen in our case report. Reports from other centres in Nigeria also corroborated this assertion (10-11).

Our case (Baby OB) typifies majority of the risk factors that has been recognised to predispose a jaundiced newborn to bilirubin encephalopathy. These include preterm, delivery outside the hospital, late presentation and G-6-PD deficiency as equally observed by other workers (10-11). The fact that there was a similar occurrence in the 30 year old mother of the baby manifesting now with features of cerebral palsy made the condition more appalling and of great interest. This underscores the reality that not much progress has been made in our attempt at reducing the burden of newborn brain damage from severe neonatal jaundice in Nigeria as corroborated by past and recent reports from our centre and similar centres in Nigeria (5, 7-11). We report this case to refocus attention on the silent killer of newborns and a major cause of handicap of children in Nigeria.

Ignorance, poor knowledge about jaundice by mothers and health workers alike has been widely reported to contribute to late presentation of babies with jaundice and subsequent development of bilirubin-induced brain damage (10-13). Other contributory factors include delivery outside a health facility and absence of home visit (10-13) as seen in our case report.

Caregivers often do not appreciate jaundice especially in dark skin newborns and primary

health care workers are often confused on whether to refer a jaundice newborn or just to reassure the mother (13). They often do not have specific guidelines to follow. Though there have been publications highlighting what may constitute danger signs in jaundiced newborns (11, 13), locally adaptable guidelines particularly for primary health care workers in Nigeria will be worthwhile.

The American Academic of Paediatrics (AAP) (14) proposed a guideline to assist clinicians in recognition and management of newborn with jaundice. The overall aim of this guideline is to promote an approach that will reduce the frequency of severe neonatal hyperbilirubinaemia and bilirubin encephalopathy and minimize the risk of unintended harm such as increased anxiety, decreased breastfeeding, or unnecessary treatment for the general population and excessive cost and waste. These guidelines emphasize the importance of universal systematic assessment for the risk of severe hyperbilirubinaemia, close follow-up and prompt intervention when indicated.

We advocate increased awareness creation not only to mothers and potential mothers but also to health workers, religious leaders, market women, traditional birth attendants and indeed all stakeholders. In addition, recognition and appropriate counselling of mothers and potential mothers should be part of the Integrated Management of Maternal, Newborn and Child Health Strategy (IMNCS). Compulsory home visit by health care workers to newborn at least once during the first week of life should be routine. This will not only help with early recognition and treatment of newborn jaundice, it will also help in the control of other neonatal conditions like sepsis and tetanus. Furthermore, specific and practicable instructions about the prevention, recognition and emergency nature of neonatal jaundice should be taught in schools as part of the School Health Programme to inform and instruct the leaders and parents of tomorrow on the need to join the fight to eliminate an important terminator and destroyer of our children future.

## Conclusion

Kernicterus is a significant cause of morbidity and mortality in Nigerian children. It is often neglected and not afforded the attention it deserves. We advocate more aggressive preventive measures and involvement of all stakeholders to nip this problem in the bud.

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## Authors' contribution

KBK conceive the idea, took part in the management of the patient, the drafting and review of the manuscript, OT, AAT and KDK took part in the management of the patient and the drafting and review of the manuscript. All the authors approved the final manuscript.

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