East African Medical Journal Vol. 95 No. 10 October 2018
CEREBRAL PALSY DUE TO KERNICTERUS - CAN WE DO MORE FOR OUR PATIENTS?
Kevin Tony Okoth, Maseno University, School of Medicine, Kisumu, Kenya, P.O. Box 167, Kendu, Bay. Denis Kiplangat McKnight, Maseno University, School of Medicine, Kisumu, Kenya. Brenda Kanali Luseno, Maseno University, School of Medicine, Kisumu, Kenya, P.O. Box 3148, Nakuru. Medical Students, Maseno University, School of Medicine, Kisumu, Kenya.

Corresponding author**:** Kevin Tony Okoth, Maseno University, School of Medicine, Kisumu, Kenya, P.O. Box 167, Kendu Bay. Email: kevintonyokoth@gmail.com

CEREBRAL PALSY DUE TO KERNICTERUS - CAN WE DO MORE FOR OUR PATIENTS?

K. T. Okoth, D. K. McKnight and B. K. Luseno

ABSTRACT

Kernicterus is still a health concern in the pediatric age group. Has a high morbidity and mortality rate? This report highlights a case study of a child with kernicterus. The patient is a 1 year two-month-old male, with history of prematurity, asthma, and HEI currently on septrin and seronegative, presented with a two-day history of convulsive episodes with fever, choreoathetosis head movements and delayed milestones.

INTRODUCTION

Cerebral palsy is characterized by non-progressive impairment of the motor system, as a result of brain lesion and anomalies. These can arise before birth, or after.[1] It has been postulated to be the most common cause of motor disability in childhood, with an estimated population of 1.5-4 per 1000 live births.[2] A systematic review of cerebral palsy in Africa showed that prevalence varied from 2-10 per 1000 live births. Birth asphyxia, preterm birth, kernicterus, low birth weight, and CNS infections were established to be the most common etiologies.[3]

Kernicterus is the deposition of bilirubin in the brain, might be the cortex or the brain ganglia. Patients present differently and to a varying extent depending on the location and severity of the lesion. There is no clear-cut cause-effect relationship between kernicterus and the degree of hyperbilirubinemia. However, laboratory and clinical evaluation of patients have determined that kernicterus is neurotoxic.[4] Hyperbilirubinemia affects the basal ganglia, substantia nigra, hippocampus and the brainstem nuclei. Clinical presentations depict the characteristic pattern of neuronal necrosis. Population surveys demonstrate that kernicterus occurs when hyperbilirubinemia exceeds 25mg/dL, and this is more in the Hispanic and Asian populations. Male infants have a higher risk of developing hyperbilirubinemia, and the symptoms manifest in the first few days of life.[4]

CASE PRESENTATION

A 1year 2-month-old male, who is HIV exposed, presented with Convulsions lasted for 5 minutes, came at 30-minute intervals in clusters of 2-4 episodes for the past two days. Generalized tonic-clonic in nature. No identifiable pre-ictal symptoms. Ictal phase involved eye-rolling, lip-smacking, frothing, loss of bowel and bladder control, and shaking movement of upper and lower limbs.

There was associated hotness of body, passing of loose stool, and cough with rhinorrhea three days before convulsions. In the review of systems, No history of yellowness of the eyes, abdominal distension, the passing of dark or blood in urine, no history of head trauma. There was no difficulty in breathing or vomiting. Mother noticed purposeless shaking movements of the head around eight months of age. The patient is known asthmatic and was diagnosed at eight months of age, and the asthma is well controlled by salbutamol. He has had a convulsion at six months of age and is maintained on phenobarbital. Patient, who is seronegative and on septrin prophylaxis, was born to a sero-reactive mother on Highly Active Anti-Retroviral Therapy (HAART).

The mother attended three antenatal visits and was not managed for any illnesses, but was on HAART. Mother reports giving birth at five months' gestation, but this was questionable due to the possibility of wrong dates. Delivery was via caesarian section secondary to antepartum hemorrhage. The outcome was a live male infant, weighing 1.6kg, resuscitated, and admitted at the newborn unit for two weeks before the mother was able to see him. At the time of presentation, the patient had strong neck support, sitting without support by eight months, not crawling or walking but cooing and has never talked. Immunization was up to date, and the child was exclusively breastfed up to 7 months. Currently, he is still on expressed breast milk.

Physical examination revealed a male child who was in general fair condition, not in pain or respiratory distress, well-nourished and not dehydrated, jaundiced, cyanosed, or edematous. The weight was 6.7kg, heart rate at 130bpm, respiratory rate at 46bpm, saturating at 96% on room air and a fever of 39 degrees Celsius. The respiratory, abdominal, cardiovascular and head, ear, nose, and throat exam were normal. CNS examination findings were an alert child, with both cranial nerves grossly intact, muscle bulk and tone were normal. The patient could not bear weight without support, but muscle power appeared normal. He also had non-rhythmic, purposeless, involuntary shaking movement of the head. Kerning's and Brudzinski signs were negative.

Lumbar puncture and CSF for biochemistry, culture, and acid-fast bacilli turned normal. Renal function tests were normal. Full hemogram showed mild anemia (Hb 9.9g/dl), hypochromic microcytic with no WBC change. Blood smear for malaria parasites showed no parasites. RBS was 3.9mmol/l. An impression of delayed developmental milestones in a child with choreoathetosis movements consistent with cerebral palsy was made. Treatment for meningitis was initiated using ceftriaxone and patient maintained and discharged on the same dose of phenobarbital. C.T. brain without contrast was healthy, and an EEG was prescribed. The patient was referred to physiotherapy and occupational therapy and is to be followed up in the medical outpatient clinic.

DISCUSSION

This case study draws attention to the need for early diagnosis of neonatal jaundice. After diagnosis, prompt and adequate actions need to be taken to avert the long-term sequela as seen in the patient we presented. The main issue of concern is how to identify the high-risk mothers and infants, how to pool affordable resources in diagnosing and treating, and finally, how to continue long term management of such patients. One of the identifiable risk factors for this patient was low birth weight and prematurity, which is among those described by. [3] Identifying high-risk mothers require a thorough antenatal evaluation for risk factors like antepartum hemorrhage, multiple gestations, intrauterine pathology, intrauterine growth restriction, ABO and rhesus incompatibility. A cross-sectional study from Iran identified the relationship between the mother's white blood cells, platelets, hemoglobin, and gestational age with neonatal jaundice.[5]

Transcutaneous bilirubin meter provides bilirubin levels as compared[6] to visual inspection and requires fewer blood tests than visual assessment for similar results, hence is suitable for identifying high risk infants. Early manifestations of Acute Bilirubin Encephalopathy include reduced alertness, hypotonia, poor feeding, and weak Moro reflex.[7] Affordable screening equipment has been identified, like the BiliSpec pilot study in Malawi.[8] Additionally, prompt treatment by phototherapy or exchange transfusion, as guided by age appropriated bilirubin levels,[1,2] improves outcome in these infants. For those who have already developed cerebral palsy, physiotherapy, speech therapy, and occupational therapy should be initiated as soon as possible to improve their quality of life, and to avoid long term complications of immobility and impaired speech.[9]

CONCLUSION

Kernicterus is a preventable condition that can be arrested at the early stages of neonatal jaundice. Health practitioners need to have a high index of suspicion in identifying the situation and promptly managing it. For patients who present late, or in whom the disease has progressed, there is a need for referral for further management at physiotherapy, speech therapy, and occupational therapy departments. People with cerebral palsy also deserve a better quality of life, which is only possible through a multidisciplinary approach.

REFERENCES

1. Marcdante K, Kliegman R, Nelson W. Nelson essentials of pediatrics. Philadelphia, Pa: Elsevier Saunders; 2015.
2. Christensen D, Van Naarden Braun K, Doernberg N, Maenner M, Arneson C, Durkin M et al. Prevalence of cerebral palsy, co-occurring autism spectrum disorders, and motor functioning - Autism and Developmental Disabilities Monitoring Network, USA, 2008. Developmental Medicine & Child Neurology. 2013;56(1):59-65.
3. Karapurkar T, Murphy C, Schendel D. The prevalence of developmental disabilities among multiple births in Metropolitan Atlanta. Paediatric and Perinatal Epidemiology. 2008;15(4):A18-A18.
4. Springer S, Annibale D. Kernicterus Clinical Presentation: History, Physical, Causes [Internet]. Emedicine.medscape.com. 2014 [cited 25 September 2019]. Available from: https://emedicine.medscape.com/article/975276-clinical#b8
5. Mojtahedi S, Izadi A, Seirafi G, Khedmat L, Tavakolizadeh R. Risk Factors Associated

with Neonatal Jaundice: A Cross-Sectional Study from Iran. Open Access Macedonian Journal of Medical Sciences. 2018;6.

6. Kaplan M, Shchors I, Algur N, Bromiker R, Schimmel M, Hammerman C. Visual screening versus transcutaneous bilirubinometry for predischarge jaundice assessment. Acta Paediatrica. 2008;97(6):759-763.
7. Kaplan M, Bromiker R, Hammerman C. Hyperbilirubinemia, hemolysis, and increased bilirubin neurotoxicity. Seminars in Perinatology. 2014;38(7):429-437.
8. Keahey P, Simeral M, Schroder K, Bond M, Mtenthaonnga P, Miros R et al. Point-of-care device to diagnose and monitor neonatal jaundice in low-resource settings. Proceedings of the National Academy of Sciences. 2017;114(51):E10965-E10971.
9. Ganesh G, Das S. Evidence-based approach to physical therapy in cerebral palsy. Indian Journal of Orthopaedics. 2019;53(1):20.