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DOI: http://ijmer.in.doi./2021/10.08.78

INTERNATIONAL JOURNAL OF MULTIDISCIPLINARY EDUCATIONAL RESEARCH
ISSN:2277-7881; IMPACT FACTOR :7.816(2021); IC VALUE:5.16; ISI VALUE:2.286
Peer Reviewed and Refereed Journal: VOLUME:10, ISSUE:8(4), August: 2021
Online Copy of Article Publication Available: www.ijmer.in
Digital certificate of publication: http://ijmer.in/pdf/e-Certificate%20of%20Publication-IJMER.pdf
Scopus Review ID: A2B96D3ACF3FEA2A
Article Received: 4th August
Publication Date:30th August 2021

BILIRUBINEMIA LEADING PROGRESSIVE SENSORINEURAL HEARING LOSS- A CASE STUDY

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Abstract

Hyperbilirubinemia is a common neo natal problem with toxic effects on the nervous system that can cause hearing impairment. This article is based on a case of progressive sensorineural hearing loss. Hearing impairment occurs about 10-50 times more frequently in neonates with severe jaundice. Total bilirubin level has the highest predictability for infant hearing status. Blood group and Rhesus (Rh) incompatibilities between mother and child and G6PD deficiency are important known cause for hearing impairment due to jaundice. Childhood jaundice is included in the High-Risk Register by Joint Committee of Infant Hearing. In this case we observed how neonatal jaundice can cause a progressive sensorineural hearing loss if early diagnosis was not carried out.

Keywords: Hearing Loss, Jaundice, Newborn Risk Factor, Sensorineural.

Introduction

After 30 days of neonatal life babies are mostly hospitalized for Jaundice (1, 2). However, the effect is mild and cured within a fortnight. Bilirubin level exceeding beyond the 95% percentile in 8-11% of newborns necessitates further examination and treatment (2), otherwise serious complications like kernicterus can cause lifelong disabilities (3,4). Bilirubin-induced lifelong neurological conditions can also occur in some infants. Early diagnosis and treatment are that the key to tackle the future effect of bilirubin in infants. Despite recent progress in jaundice care, bilirubin toxicity remains one of the main challenges in neonatal care (5). Neonatal jaundice is one among the explanation for early sensorineural deafness in developing countries and increased blood indirect bilirubin can cross the blood-brain barrier and deposit within the auditory ventricular nucleus cells (6). Risk factor of deafness varies at different ages. A case history of deafness, congenital infections, auditory pharyngeal disorders, meningitis, ototoxic medications, and bilirubin levels above 20 mg/dl are involved in deafness in children under 2.5 years old (7,8). Since early detection and treatment of deafness during the primary 6 months of infancy has the simplest impact on language development, performing infantile screening tests for detection of deafness is additionally necessary(9,10).The Otoacoustic emission (OAE) test is one of the routine after birth screening procedures for risk factor free infants (11, 12).The auditory brainstem response (ABR) is another test used to detect deafness and neural type especially (11), and has the efficiency and sensitivity required for infants with hyperbilirubinemia (12, 13). Several studies have investigated the connection between hyperbilirubinemia and hearing impairments. Kernicterus has been reported because the commonest explanation for acquired deafness (13), and therefore the risk of deafness has been shown to extend with prolonged exposure to hyperbilirubemia.

Case-Study

A case of 1 years old girl was referred for hearing assessment and speech-language evaluation. As per the parent's information, the client was born as per normal delivery and birth-cry was present. On 3rd day client was diagnosed with high-level neonatal jaundice (Hyperbilirubinemia) that she was admitted to the NICU for 8 days and a transfusion was done. After 1month, the mother observed that client isn't responding even to loud sounds then the parents consulted to ENT specialist and Audiologist. At the age of 1 year during her consultation with the Audiologist firstly we examine the Tympanic membrane by otoscopic examination and observed a normal intact tympanic membrane. Oto Acoustic Emission (OAE) test was carried out. OAE test especially distortion product OAE (DP OAE) generally used to detect the function of inner hair cells. This test is mostly used for screening purposes and in this case, the result of DP OAE was "refer" which is indicative of a sensorineural or conductive hearing loss. To rule out conductive hearing loss tympanometry and acoustic reflex threshold are calculated. The tympanometry result shows "A" type tympanogram and reflex present at 500 Hz, 1000 Hz, and 2000 Hz frequencies in both ears which are indicative of a normal middle ear function. This result leads us to be confirmed that the child has sensorineural hearing loss. To detect the site of lesion we have conducted Brainstem Evoked Response Audiometry (BERA) test by using two-channel recording electrode montages. Before starting the test, we used sedation for a deep sleep of the patient. We started giving click stimulus to the patient at a sound level of 100 dBnHL and found presence of wave V at 7.3 msec time window but the wave V was absent when we tested the ear at 80 dBnHL but preset at 85 dBnHL. We again reevaluated the BERA test for repeatability check and found the same results which indicated the child has a moderately severe to severe hearing loss. After the assessment procedure we counseled the parents about the problems and suggested using a hearing aid to maintain her residual hearing, and for a follow-up on an annual basis.

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At age of 2- ½ years, the child's hearing level was re-evaluated by using an informal hearing assessment tool that is Play audiometry. During this evaluation procedure, we first habituated the patient with the sound and then began the test procedure in a sound treated chamber by using MAICO MA 42 dual channel audiometer. The sound was presented at 110 dB HL and by using the Modified Hugshon-Wastelake procedure we calculated the hearing threshold at each frequency respectively. Here we evaluated the pure tone average of both ears and found the child has bilateral severe sensorineural hearing loss. After completing the informal assessment, we move to conduct BERA test. Before conducting BERA test sedition was given to the child and the test was administered by using dual channel electrode montages in a sound treated chamber. The sound was presented at 100 dBnHL by headphones and we observed a wave V at 6.9 msec time epoch. Again, we reduced the sound intensity and presented at 90dBnHL and 85dBnHL. In both intensities wave V was calculated in which wave V was present at 7.4msec time but wave V absent at 85dBnHL sound intensity. This indicated a severe to profound hearing sensitivity of the patient. We used same instruments same condition in both of the testing. From the BERA test, we observed a progressive loss of hearing in the patient. With regular use of hearing aid and continue speech language stimulation –therapy, the client can enroll in teacher's college (class 2 in Kendriya Vidyalay). As an explanation for deafness in client was neonatal jaundice, NICU, her deafness is progressive in nature but with early identification and intervention, client can enroll in normal speech hearing environment.

Discussion and conclusion

As per this case and review, the danger of hearing impairment in icteric infants with a bilirubin level above 20mg/dl was 10-50 times higher. Sensorineural deafness finds to be associated with severity of hyperbilirubinemia, explanation for jaundice and treatment approach. An increased rate of complications including microcephaly, abnormal eye movements and developmental delay was seen in icteric infants with deafness. While other studies have reported a various range of incidence rates for abnormal ABR test results among infants with hyperbilirubinemia, showed an incidence rate of 4.8%. However, hyperbilirubinemia required blood transfusion can damage the hearing system and disrupt hearing tests even without developing kernicterus. The mechanism underlying bilirubin neurotoxicity hasn't been elucidated. Also, it's not clear how just some of the infants with a particular level of bilirubin develop deafness or neurological damage. To prevent this high-level hearing loss early identification of hearing loss is very much necessary. According to Joint Committee on Infant Hearing (2007) has provided High Risk Register criteria for conducting compulsory hearing screening at the age of 1 month prior the birth of the child.

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