

Original Research Article

Screening of infants and young children with hyperbilirubinemia for sensorineural hearing impairment: an institutional study

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ABSTRACT

Background: Jaundice is one of the most common problems occurring in newborns. Severe neonatal hyperbilirubinemia is a common cause of sensory neural hearing loss. It is important to identify and treat the jaundice early, to prevent complications like encephalopathy leading to hearing loss. Early detection of hearing loss is important for early intervention. The aim of the study was to evaluate the prevalence of sensorineural hearing loss (SNHL) in children with hyperbilirubinemia and to correlate the degree of hearing loss with degree of hyperbilirubinemia.

Methods: The 50 cases including newborn and young children (<12 years of age) having history of hyperbilirubinemia and exchange transfusion were included in this study. After the approval and clearance from institutional ethical committee, this study was conducted from April 2021 to March 2022 in the department of ENT, govt. medical college Amritsar in co-ordination with department of pediatrics.

Results: On comparing brain stem evoked response audiometry (BERA) and otoacoustic emissions (OAE) results, it was found that out of 50 patients BERA was abnormal in 8 patients while 42 had normal BERA. 18% (9) patients had abnormal OAE results out of which 7 had bilateral REFER result while 2 had unilateral REFER result. On comparing the OAE and BERA for sensitivity and specificity, the sensitivity was 92.9% in right ear while specificity was 62.5%, whereas in left ear sensitivity was 95.2% while specificity was 75%.

Conclusions: According to our study the prevalence of sensorineural hearing impairment comes out to be 16% as per BERA. BERA is more sensitive and specific as compared to OAE. But still OAE can be used as screening test considering the cost factor and ease of conducting OAE test while BERA is a confirmatory test for SNHL.

Keywords: Hyperbilirubinemia, OAE, BERA, Acute bilirubin encephalopathy, Neonatal jaundice, Direct venous exchange transfusion

INTRODUCTION

The auditory pathway is one of the central nervous system's most vulnerable pathways to noxious chemicals. SNHL and auditory neuropathy are mostly caused by severe newborn hyperbilirubinemia. SNHL is a type of hearing loss due to pathology of the cochlea, auditory nerve, or central nervous system or in other words, SNHL is a result of damage to the hair cells within the inner ear,

the vestibulocochlear nerve, or the brain's processing centre. This is different from conductive hearing loss, which results from the inability of sound waves to reach the inner ear.

The incidence of SNHL is different in different countries. In developed countries like United States, incidence of sudden SNHL is between 5-27 per 100,000 people each year, with approximately 66,000 new annual cases.¹ As

per the statistics of national programme for the prevention and control of deafness (NPPCD) given to WHO, the estimated prevalence is 6.3% in Indian population.²

SNHL can occur at birth (congenital) or can be acquired over time. Hyperbilirubinemia, toxoplasmosis infection in mother, rubella, cytomegalovirus, herpes, or syphilis, genetic factors and syndromes in some children at birth, low birth weight and hereditary causes are few of the known aetiologies for congenital SNHL in newborn.³

Jaundice is also known as hyperbilirubinemia. In babies, hyperbilirubinemia is independent risk factor for SNHL.⁴

BERA and OAE are recommended for screening of hearing in newborns. Both BERA and OAE techniques are inexpensive, automated, and reproducible. OAEs are frequency dispersive responses arising from the cochlea. Since OAE evaluates the hearing from middle ear to the outer hair cells of inner ear, it is used to screen for SNHL. Whereas BERA is an electrophysiological measurement that is used to assess auditory function from 8th nerve to the inferior colliculus of midbrain in response to a click stimulus. OAE is an ideal tool for screening whereas BERA is a gold tool for confirmation of hearing loss.⁵

METHODS

The present study entitled “screening of infants and young children with hyperbilirubinemia for sensorineural hearing impairment: An institutional study” was done in department of ENT, govt. medical college, Amritsar.

Inclusion criteria

Newborn with gestational age of >35 weeks having hyperbilirubinaemia requiring exchange transfusion and infants and young children <12 years of age, with history of hyperbilirubinaemia requiring exchange transfusion in the neonatal period were included in study.

Exclusion criteria

Patients with congenital hearing loss, intrauterine infections, meningitis, birth asphyxia, intraventricular haemorrhage, neuro-degenerative diseases, family history of SNHL, congenital craniofacial abnormalities, dysmorphic baby, birth trauma and GA less than 35 weeks preterm babies were excluded.

All the newborn admitted in pediatrics department having hyperbilirubinemia and requiring exchange transfusion, were screened for hearing impairment. Infants and young children visiting OPD with the history of hyperbilirubinemia requiring exchange transfusion in the neonatal period were also included in the study. All the enrolled cases were subjected to detailed history regarding clinical course, neurological examination (to evaluate the bilirubin encephalopathy) and complete

general physical examination along with ENT examination with special reference to ear.

All the newborn were subjected to initial screening with OAE before discharge. It was followed by screening with BERA after three months and the interpretations were recorded. All young children with <12 years of age with history of DVET (Direct venous exchange transfusion) for NNJ (Neonatal jaundice), visiting OPD subjected to OAE initially and BERA was done on appointment.

RESULTS

On distributing the patients according to their gender, out of total 50 cases 58% were male and 42% were female patients. Out of these 50 cases 54% had birth weight of 2.5 kg to 4 kg while only 6% patients had birth weight less than 1.5 kg (Table 1).

Table 1: Distribution of patients according to birth weight along with chi-square test results.

Birth weight (kg)	N	Percent (%)	Chi-square
1.49 and less	3	6.0	X ² =18.280 p=0.001
1.5-2.49	20	40.0	
2.5-4	27	54.0	

On comparing gestational age, 58% patients had 35-36 weeks, 30% had GA of 37-40 weeks while only 12% of patients had 41 weeks and above (Table 2).

Table 2: Distribution of patients according to gestational age along with chi-square test result.

Gestational age (week)	N	Percent (%)	Chi-square
35-36	29	58.0	X ² =16.120 p=0.001
37-40	15	30.0	
41 and above	6	12.0	

Distributing patients in groups it was observed that 68% patients had bilirubin levels of 16-23 mg/dl, 26% having peak bilirubin levels of 24-31 mg/dl while 6% patients had peak bilirubin levels of 32-38 mg/dl (Table 3).

Table 3: distribution of patients according to peak bilirubin level along with chi-square test result.

Peak bilirubin level (mg/dl)	N	Percent (%)	Chi-square
16-23	34	68.0	X ² =51.440 p=0.001
24-31	13	26.0	
32-38	3	6.0	

Early onset sepsis was major cause of hyperbilirubinemia in 46% patients followed by ABO incompatibility 34% and Rh incompatibility in 20% patients (Table 4).

Table 4: Distribution of patients according to cause of jaundice along with chi-square test result.

Cause of jaundice	N	Percent (%)	Chi-square
ABO incompatibility	17	34.0	X ² =5.082 p=0.07
Rh incompatibility	10	20.0	
Early onset sepsis	23	46.0	

All patients were subjected to OAE screening and BERA test for hearing assessment. OAE screening showed that 7 patients had bilateral REFER results and 1 patient had REFER result in right ear and 1 in left ear (Table 5).

Table 5: Distribution of patients according to OAE results.

OAE	Right ear	Left ear
REFER	8	8
PASS	42	42

BERA was found to be abnormal in 8 patients while 42 had normal BERA (Table 6). Out of these 8 abnormal BERA, 6 had REFER results on OAE while 2 had PASS results on OAE. And out of 42 normal BERA, 3 patients had REFER results on OAE and 39 had PASS results. Among the 8 abnormal BERA patients, 6 had sensorineural hearing loss while 2 were diagnosed with Auditory neuropathy spectrum disorder (ANS), in these 2 patients OAE results were PASS while BERA was abnormal. Result in right ear and one in left ear.

Table 6: Distribution of patients according to BERA results along with chi-square test result.

BERA results	N	Percent (%)	Chi-square
Normal	42	84.0	X ² =23.120 p=0.001
Abnormal	8	16.0	

It was observed that out of 34 patients with peak bilirubin levels of 16-23 mg/dl OAE results were abnormal in 7 patients on initial screening while 4 showed abnormal results on BERA. In patients with peak bilirubin level of 24-31 mg/dl OAE and BERA results was abnormal in 1 patient while all the 3 patients with peak bilirubin level of 32-38 mg/dl showed abnormal BERA results (Table 7).

Table 7: correlation of peak bilirubin levels with results of BERA and OAE.

Peak bilirubin levels (mg/dl)	N	Abnormal OAE	Abnormal BERA
16-23	34	7	4
24-31	13	1	1
32-38	3	1	3

On comparing the OAE and BERA for sensitivity and specificity, in right ear the sensitivity was 92.9% while

specificity was 62.5%, whereas in left ear sensitivity was 95.2% while specificity was 75%.

DISCUSSION

As per the study done by Meredith porter et al physiologic jaundice in healthy term newborns follow a typical pattern.⁶ The average total serum bilirubin level usually peaks at 5 to 6 mg/dl (86 to 103 μ mol per Lt.) on the third to fourth day of life and then declines over the first week after birth. In the present study majority of the babies (68%) had peak bilirubin levels in the range of 16-23 mg/dl. This was followed by 24-31 mg/dl in 26% and 32-38 mg/dl range in 6% of cases. Out of these patients, signs of bilirubin encephalopathy were seen in 3 patients with serum bilirubin levels range of 16-23 mg/dl.

In our study early onset sepsis (46%) was the most common cause of jaundice followed by ABO incompatibility (34%) and Rh incompatibility (20%). In a study done by Kaplan et al ABO blood group hetero-specific newborns (mother group O, new-born A or B) were found to be at risk of hyperbilirubinemia due to immune based hemolysis.⁷

All the study participants underwent OAE screening test and 14% (7) participants had bilateral abnormal OAE results while 4% (2) had unilateral abnormal OAE (1 REFER in right ear and 1 REFER in left ear). Hence 18% (9) patients had OAE abnormality among the total study population. A study done by Baradaranfar et al showed that, out of 35 children with hyperbilirubinemia, 14.3% had REFER results while 85.7% had PASS results.⁸

Among the study participants, 16% (8) patients had abnormal BERA results while BERA was normal in 84% (42) patients. Results of our study were consistent with a study done by Mirajkar et al in which out of fifty eligible neonates, eight (16%) had hearing impairment, as per BERA analysis, twenty-one (42%) had hearing impairment as per OAE analysis.⁹

In our study when patients were distributed according to the peak bilirubin levels, out of 34 patients, with bilirubin level of 16-23 mg/dl 7 patients had abnormal OAE results while 4 patients had abnormal BERA results. While all 3 patients with peak bilirubin level of 32-38 mg/dl had abnormal BERA results.

Our results were consistent with study by Wickremasinghe et al in which the risk of SNHL in babies with bilirubin levels at or above the American academy of pediatrics exchange transfusion limits (ETT) was quantified. Only bilirubin levels 10 mg/dl higher than ETT were linked with a statistically significant increased risk of SNHL. Similarly, only bilirubin levels of 35 mg/dl were linked with an elevated risk of SNHL.¹⁰

In another study conducted by Ullah et al in hospital of Islamic Azad university, Iran in 97 newborns who were

hospitalized it was reported that the rate of hearing impairment was higher in high risk population of newborn with severe pathological hyperbilirubinemia. Higher the bilirubin concentration, higher was the risk of impairment to auditory system.¹¹

In a prospective study done by Bhatt et al to assess the sensitivity and specificity of OAE test in newborns comparing with auditory brain stem response (ABR) The sensitivity and specificity of OAE was 70% and 61% at 0 month and 70% and 99% at 3 month and BERA sensitivity and specificity at 3 month was 90% and 99% and at 6 month 100% and 99% respectively. In our study on comparing the BERA and OAE for sensitivity and specificity, in right ear the sensitivity was 92.9% while specificity was 62.5%, whereas in left ear sensitivity was 95.2% while specificity was 75%.¹²

CONCLUSION

Based on our study, the prevalence of sensorineural hearing impairment in hyperbilirubinemia is estimated to be 16% as determined by BERA. While there is a clear correlation between high bilirubin levels and hearing impairment, our findings suggest that other factors may also play a role in the development of hearing impairment in these cases. We recommend further studies with a larger number of cases and follow-up BERA assessments at 6 months and 1 year of age to better understand the long-term effects. In terms of screening for hearing impairment, our study indicates that BERA is more sensitive and specific compared to OAE. However, considering the cost-effectiveness and ease of conducting OAE, we suggest using OAE as a screening test. BERA should then be used as a confirmatory test for hearing assessment in cases where OAE results are inconclusive or where further evaluation is warranted.

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Conflict of interest: None declared

Ethical approval: The study was approved by the Institutional Ethics Committee

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