



Supra-Threshold Hearing Sensitivity Disorders and Mild Permanent Hearing Loss: Neglected Cause of Hidden Hearing Loss and Speech Defects

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Abstract Supra-threshold disorders in the form of auditory neuropathy (AN)/auditory dys-synchrony (AD) or central auditory processing disorders (CAPD), a special type of retrocochlear hearing loss; and also children with mild permanent hearing loss (PHL), may be missed on current hearing protocols. Otoacoustic emissions (OAE) and Brain stem evoked response audiometry (BERA), are tests, when used in combination, can indirectly help diagnose the different types of hearing loss. To correlate the parental awareness and the pattern of hearing loss (HL) in children with/without auditory and/or speech problems, using both OAE and BERA as hearing tests for indirect identification of suprathreshold disorders and mild PHL. An Observational Prospective study carried out on 100 children in the age group of 0–5 years, for detection of type of HL by both OAE and BERA and correlating it with parental awareness of HL and speech defects. In 72.22% of children with speech problem only and without any complaints of HL, some form of HL (confirmed HL—OAE refer/BERA fail or auditory neuropathy/auditory dys-synchrony—OAE pass/BERA fail) was diagnosed, whereas 24.07% had no detectable HL on both OAE and BERA pass, and were considered as indirect evidence of CAPD. 3.7% with OAE refer and BERA pass were considered indicative of mild PHL. AN/AD/CAPD/Mild PHL are important cause of speech delay without parental awareness of HL. OAE and

BERA together can be used as an indirect evidence of their presence.

Keywords Retrocochlear hearing loss · Otoacoustic emissions · Auditory brain stem evoked responses · Brainstem auditory evoked potential · Hearing loss · Speech disorder · Auditory neuropathy · Central auditory processing disorders

Introduction

Hearing loss in children constitutes a considerable handicap because it is an invisible disability and compromises optimal and personal achievement of a child. If the diagnosis is delayed by 24–36 months of age, after this age even rehabilitation procedures (like hearing aids, cochlear implant, speech therapy, psychological intervention on family) are unable to ensure complete development of speech, thus preventing the full participation of deaf child in social living. Pediatric hearing loss (HL) is a major concern in India due to high incidence of HL in this age, and lack of facilities and awareness for early diagnosis, in proportion to the population size.

5% of Indian population have speech and hearing problem due to congenital sensorineural hearing (SNHL) with delayed development of speech and language (DOSL) [1]. If a child has hearing impairment, early detection followed by appropriate treatment provides the best chance for maximizing the critical period of hearing, to improve hearing and oral communication skills. On the other hand, late detection and treatment leave the child with poor speech development and school achievement [2–4]. Otoacoustic emissions (OAE) and brain stem-evoked response audiometry (BERA), are two common tests for

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audiological assessment after birth. The latter is also known as auditory brainstem response (ABR)/brain stem auditory evoked response (BSAER). Suprathreshold disorders are a type of hearing impairment, that may or may not be associated with hearing sensitivity loss on audiogram [5]. The disorders under this category are auditory neuropathy/auditory dys-synchrony (AN/AD) and central auditory processing disorders (CAPD). This entity has been used to describe disorders of auditory processing, with hearing ability out of proportion to the degree of hearing sensitivity loss on audiogram. These conditions may be diagnosed on electrophysiologic tests like BERA, Mid-latency response (MLR) and Late latency response (LLR). Cochlear hearing loss is characterized by loss of OAE, whereas both cochleogenic and neurogenic auditory neuropathy (AN) have preservation of OAE due to cochlear echoes, being generated by outer hair cells, but absent waves on BERA [5].

In countries, with high patient burden and unmotivated population, universal hearing screening programmes are not feasible. High risk screening is mostly advocated. However, in rural centres, even this is not being practiced due to the ignorance on part of parents and the peripheral health workers. Most of the children present late, after the critical period of hearing, with resultant inadequate hearing and oral communication skills. In 2006, India launched the National Programme for Prevention and Control of Deafness, which aims to identify babies with bilateral severe-profound hearing losses by 6 months of age and initiate rehabilitation by 9 months of age [6]. Under this programme, a three- step protocol of OAE followed by BERA, is being followed in Institution-based screening—to screen every baby born in a hospital or admitted there soon after birth using OAE. Those who fail the test are re-tested after 1 month. Those who fail the second screening are referred for BERA testing at the tertiary-level centres [6]. This protocol and also two- step protocol suggested by few, may miss many children with supra-threshold disorders in the form of auditory neuropathy/auditory dys-synchrony (AN/AD) and central auditory processing disorders (CAPD) as they may have low or no hearing sensitivity loss and present with speech problems only [7]. They pass OAE and BERA testing and hence are sent home. These children present late, when speech delay becomes evident. Some others, even with profound hearing loss and speech delay present late.

The current study was undertaken to study the role of both OAE and BERA done simultaneously, as audiological tests for indirect identification of suprathreshold disorders [auditory neuropathy/auditory dys-synchrony (AN/AD) and central auditory processing disorders (CAPD)] and mild PHL and correlating it with the parental awareness and the pattern of hearing loss (HL) in children with/

without auditory and/or speech problems, attending a rural teaching hospital of Central India.

Materials and Methods

An Observational Prospective study was carried out at the Department of Otorhinolaryngology at rural tertiary hospital of central India. 100 children (200 ears) between 0 and 5 years of age group were included in the study from time period of October 2015–August 2017. The study was approved by the Institutional Ethics Committee, vide letter no. IEC/2015-16/1635 dated 30-09-2015.

Selection Criteria for Screening of HL

Neither Universal nor high risk screening is being strictly followed in our country, due to lack of parental motivation for hearing screening in their children, till they find hearing or speech deficits in their children. Hence, all children between 0 and 5 years, who were not previously subjected to hearing screening by electrophysiologic testing and presenting for the first time, either on their own due to hearing and/or speech problems, or referred by paediatrician or otolaryngologist based on high risk criteria or who volunteered for hearing screening of their children, were subjected to testing by both OAE and BERA, after initially undergoing Age specific Behavioral Observation Audiometry and speech audiometry tests.

Inclusion criteria

1. Children in the age group of 0–5 years, with minimum age of screening at 2 weeks after birth.
2. Children admitted to neonatal intensive care unit (NICU) for hyperbilirubinemia, prematurity, neonatal septicaemia, Apgar score below 4 at 1 min or 6 at 5 min after birth.
3. Any illness requiring hospitalization for 48 h or more in the first 4 weeks of birth.
4. Use of ototoxic drugs in paediatric/otorhinolaryngology ward.
5. Children referred by the paediatrician on the basis of suspicion of hearing loss/speech delay/otherwise in the presence of any of the high-risk factors.
6. Children attending paediatric or otorhinolaryngology OPD with any of the craniofacial anomalies of the pinna and ear canal or cleft lip/cleft palate or any recognizable syndrome, where hearing loss is a known component of a syndrome like Down's syndrome, etc.
7. Children brought by the parents on suspicion of hearing loss or speech delay.
8. Normal children coming for primary immunization, whose parents consented for participation in the study.

Exclusion criteria

1. Children with absent external auditory canal (EAC), meatal atresia, anomalies of the external ear where probe insertion was not possible.
2. Tympanic membrane perforation.
3. Chronic otitis media.
4. Newborns and children, at the time of upper respiratory tract infection, purulent ear discharge, acute suppurative otitis media, conditions of the external ear like otitis externa, impacted wax were included in the study only after the condition was treated.

In the present study, out of 100 children screened, 49 children were brought by parents with complaints related to HL and/or speech defect in their children whereas remaining 51 children who were screened, presented without any complaint related to hearing or speech, but were subjected to screening tests for HL on the basis of presence of risk factors or agreed for screening, even in the absence of risk factors.

The procedure was explained to the parents and consent was obtained. A detailed history, including perinatal history was elicited from the parents preferably from the mother. This was followed by a complete ENT examination, including otoscopic examination of the ears. If any wax, debris, or foreign bodies were found in the ear canal, they were removed. All OAEs were analyzed relative to the environmental noise-floor level. Ambient noise was compensated for during OAE testing, therefore, reducing the physiological and acoustic ambient noise which is very critical for good recordings. If the result was "Pass", then no follow up OAE was done. If result was "Refer" then repeat OAEs testing was done after cleaning the EAC. Absence of emissions with using repeat DPOAE was considered as "Refer" result. *Irrespective of "Pass" or "Refer" result, all children were tested with BERA.* The OAE test was done by distortion product otoacoustic emission (DPOAE) test. (OtoRead—Screeener, software version 7.65.01 with Thermal dot matrix line printer, Interacoustics, DK.- 5610 Assens, Denmark). The electrophysiological test of Auditory Brainstem Response (ABR) test (Neuro-MEP-4 connection, Neurosoft Ltd., Ivanovo, 153032, Russia) comprised of Click-evoked ABR testing using alternating polarity clicks as stimulus, with internal range of 110–40 dB HL (in 10 dB HL steps) and electrode impedance of < 5 dB. Monoaural auditory stimulus consisting of rarefaction clicks of 100 μ s were delivered through electrically shielded earphones at the rate of 11.1/s and the response to 2000 clicks presentation was averaged. The test results were further confirmed by age specific behavioral observation audiometry results.

Both the results of OAE refer and BERA fail were considered as confirmed HL, OAE pass and BERA fail were considered as children having Auditory Neuropathy/Auditory dyssynchrony (AN/AD), OAE refer and BERA pass were considered as children with mild permanent HL [8, 9]. OAE pass and BERA pass were considered as children with no evidence of HL, but if associated with parental observation of profound hearing loss, were considered as indirect evidence of CAPD.

Results

Baseline Characteristics

The baseline characteristics of the children screened were as follows:

1. Age distribution of children at the time of screening

In this study, of the total 100 children screened, 37% of children were between 0 and 1 year of age, followed by 23% children between 1.1 and 2 years of age with the mean age of screening of 2.08 ± 1.46 years (range of age at screening: 15 days–5 years) (Fig. 1).

2. Selection criteria for screening of children according to presenting complaints (as informed by parents)/ presence of risk factors.

In the present study, out of 100 children screened, 49 children were brought by parents with complaints related to HL and/or speech defect in their children whereas remaining 51 children who were screened, presented without any complaint related to hearing or speech, but were subjected to screening test for HL, with or without risk factors (Table 1).

All 49 (49%) children, who were brought by parents, had speech problem and 27% had speech problem only. 22% of children (19 children + 3 with suspected HL) in our study, were brought with complaints of HL.

Out of 51 children screened for HL, 43(84.31%) children were screened only on account of presence of risk factor and 8 (15.69%) children consented for participation in the study for hearing evaluation in the absence of any complaints or risk factors. One or more of the following risk factors were present, which warranted hearing screening -

- *Antenatal/Maternal risk factors* like oligohydramnios, eclampsia, multiparity, Rh incompatibility, TORCH infection, etc.,
- *Perinatal/Intranatal risk factors* like birth asphyxia, meconium stained liquor, prolonged/obstructed labour and

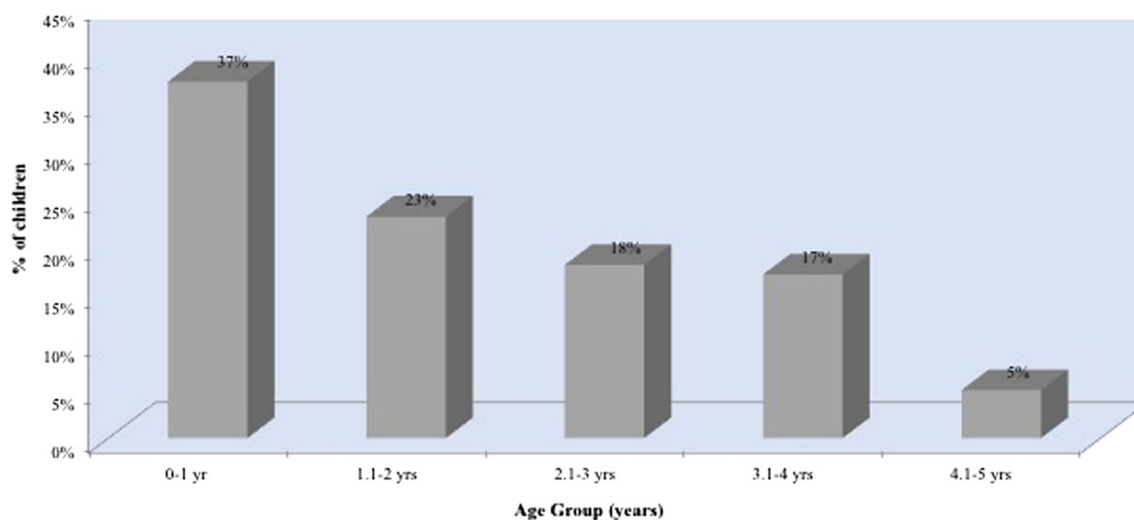


Fig. 1 Distribution of children according to the age at the day of testing

Table 1 Selection criteria for screening of children according to presenting complaints (as informed by parents)/presence of risk factors

| Presenting complaints | No. of children (%age) (N = 100) |
|---|----------------------------------|
| Profound HL with absent speech | 19 |
| Speech defect only | 27 |
| HL with speech defect | 3 |
| No complaints (N = 51) | |
| With high-risk factors present (for HL) | 43 [84.31%] |
| Without presence of high-risk factors | 8 [15.69%] |
| Total | 100 |

HL—hearing loss

- *Postnatal risk factors* like Low Birth Weight (LBW), prematurity, septicaemia, hyperbilirubinaemia, use of ototoxic drugs, syndromes/anomalies, seizure disorders, etc.

3. Distribution of speech defect/delay and HL in children (as observed by parents).

In the present study, of the total 100 children, 22% of children had both HL and speech defect/delay, as observed by parents, 27% of children were observed to have only speech defect/delay by their parents, 51% of children were not observed to have either HL or speech defect/delay. (Table 2) None of the children (0%) was brought by parents for hearing screening, on the account of observable HL only without any speech defect.

4. Distribution of associated Developmental Delay.

In the present study, out of total 100 children screened, 44% children were found to have some form of

Table 2 Distribution of speech defect/delay and HL in children (as observed by parents)

| Speech defect | Parental observation of HL (N = 100) | | Total |
|-----------------------|--------------------------------------|--------|-------|
| | Present | Absent | |
| Present | 22 | 27 | 49 |
| Absent | 0 | 51 | 51 |
| Total | 22 | 78 | 100 |
| X ² -value | 29.36, <i>p</i> = 0.0001, S | | |

HL—hearing loss

Table 3 Distribution of children according to associated developmental delay

| Developmental delay | No. of children (N = 100) | Percentage (%) |
|---------------------|---------------------------|----------------|
| Present | 44 | 44 |
| Absent | 56 | 56 |
| Total | 100 | 100 |

developmental delay (global developmental delay, motor developmental delay). (Table 3)

Pattern of Hearing Loss on OAE and BERA and Their Correlation with Observed Hearing Loss and/or Speech Defect

All the children were subjected to OAE and BERA test for identification of type of HL. The interpretation of OAE and BERA test was as follows. Both the results of OAE refer and BERA fail were considered as confirmed HL, OAE pass and BERA fail were considered as children having Auditory Neuropathy/Auditory dyssynchrony (AN/AD), OAE refer

Table 4 Pattern of hearing loss on OAE and BERA correlated with observed hearing loss and/or speech defect

| Hearing assessment | Parental observation of profound HL with absent speech) N = 19 children (38 years) No. of ears (%) | Parental observation of speech delay only (without HL) N = 27 children (54 ears) No. of ears (%) | Parental observation of both speech delay and HL | Negative History of Speech Delay and HL | Total | X2 Value | Significance |
|---------------------|--|--|--|---|-------|----------|--------------|
| OAE pass BERA pass | 1 (2.63%) | 13 (24.07%) | 0 (0%) | 28 (27.45%) | 42 | 50.101 | 0.0001, S |
| OAE refer BERA fail | 29 (76.32%) | 27 (50%) | 6 (100%) | 43 (42.16%) | 105 | 94.26 | 0.0001, S |
| OAE pass BERA fail | 7 (18.42%) | 12 (22.22%) | 0 (0%) | 31 (30.39%) | 50 | 34.85 | 0.0001, S |
| OAE refer BERA pass | 1 (2.63%) | 2 (3.70%) | 0 (0%) | 0 (0%) | 3 | 7.41 | 0.05, NS |
| Total | 38 (100%) | 54 (100%) | 6 (100%) | 102 (100%) | 200 | | |

OAE—otoacoustic emission, BERA—brainstem evoked response audiometry, HL—hearing loss

and BERA pass were considered as children with mild permanent HL [7, 8]. OAE pass and BERA pass were considered as children with no evidence of HL (Table 4, Fig. 2).

In the present study, in 38 ears of 19 children, with parental observation of profound HL and absent speech, 76.32% (29 ears) had confirmed HL, 18.42% (7 ears) had AN/AD, 2.63% (1 ear) was found to have OAE refer, BERA pass and considered as mild permanent HL (with indirect evidence of CAPD, due to profound HL, observed by parents) and 2.63% (1 ear) had no HL (again indirect evidence of CAPD). Hence, some form of HL (confirmed HL/AN), was present in 94.74% of ears in children with profound HL and absent speech. The last two situations where the ears of a same child, of 9 months of age, who had no HL detected on BERA in both the ears was

considered as an indirect evidence of central auditory processing disorder (with mild permanent HL—conductive/sensory in 1 ear).

In 54 years of 27 children, with parental observation of speech delay only (without HL), 24.07% (13 ears) had no detectable HL on OAE and BERA (*children suspected to have indirect evidence of central auditory processing disorder*), 50% (27 ears) had confirmed HL on both OAE and BERA (the parents were not aware of their hearing problem), 22.22% (12 ears) had AN/AD, that is, OAE pass but BERA fail, 3.70% (2 ears) had mild permanent HL. Hence, it was observed that 72.22% of children had some form of HL, though the children were brought by parents with complaints of speech defect/delay only and were not aware of their hearing impairment.

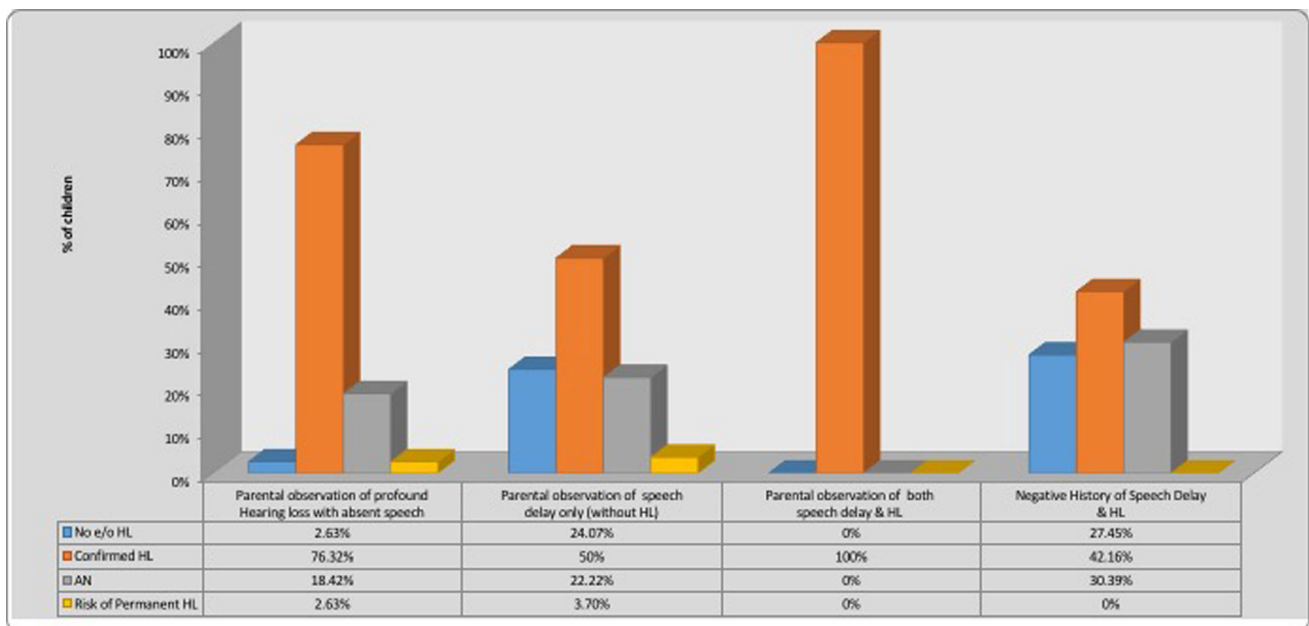


Fig. 2 Type of hearing loss by OAE and BERA correlated with parental observation of hearing loss and/or speech defect

In 6 ears (3 children) with parental observation of speech defect and HL, 100% (6 ears) showed confirmed HL on audiological testing (*positive parental history has a 100% specificity*) and there were no patients of AN/AD.

In 102 ears of 51 children with negative history of speech delay and HL, 27.45% (28 ears) had no HL on OAE and BERA testing, 42.16% (43 ears) had confirmed HL, 30.39% (31 ears) suspected AN/AD (*These were the missed children but due to screening they were diagnosed early. These included the children with presence of one or the other risk factors at birth*).

Association of Presence or Absence of Developmental Delay with Type of HL

In the present study, of the total 100 children, 44% of children had positive history of developmental delay, of which 48.86% of ears were found to have confirmed HL, 28.41% of ears showed AN/AD, 1.41% of ears were at risk

of PHL and 21.06% of ears had no e/o HL. Hence, in children with developmental delay, overall incidence of different type of HL was 77.27% (48.86% ears of confirmed HL + 28.41% ears of AN), 1.14% (1 ear) with at risk of permanent HL and 21.60% (19 ears) had no e/o HL (Table 5, Fig. 3).

In 56% of children there was no H/o associated developmental delay, of which 55.36% of ears were found to have confirmed HL, 22.32% of ears showed AN/AD, 1.78% of ears were at risk of PHL and 20.53% of ears had no e/o HL. Hence, in children without developmental delay, overall incidence of different type of HL was 77.68% (55.36% ears of confirmed HL + 22.32% ears of AN/AD), 1.78% (1 ear) with at risk of permanent HL and 20.53% (23 ears) showed no e/o HL (on comparison of overall incidence of different type of HL in children with and without developmental delay, p value = 0.70, Not Significant).

Table 5 Association of presence or absence of developmental delay with OAE and BERA screening results

| Developmental delay | No. of children (no. of ears) | Percentage (%) | OAE pass BERA pass No. of Ears (%) | OAE refer BERA fail | OAE pass BERA fail | OAE refer BERA pass |
|---------------------|-------------------------------|----------------|---|---------------------|--------------------|---------------------|
| Present | 44 (88) | 44 | 19 (21.60%) | 43 (48.86%) | 25 (28.41%) | 1 (1.14%) |
| Absent | 56 (112) | 56 | 23 (20.53%) | 62 (55.36%) | 25 (22.32%) | 2 (1.78%) |
| Total | 100(200) | 100 | 42(21%) | 105(52.5%) | 50(25%) | 3(1.5%) |
| X2-value | | | 1.42, p value = 0.70, Not significant | | | |

OAE—otoacoustic emission, BERA—brainstem evoked response audiometry

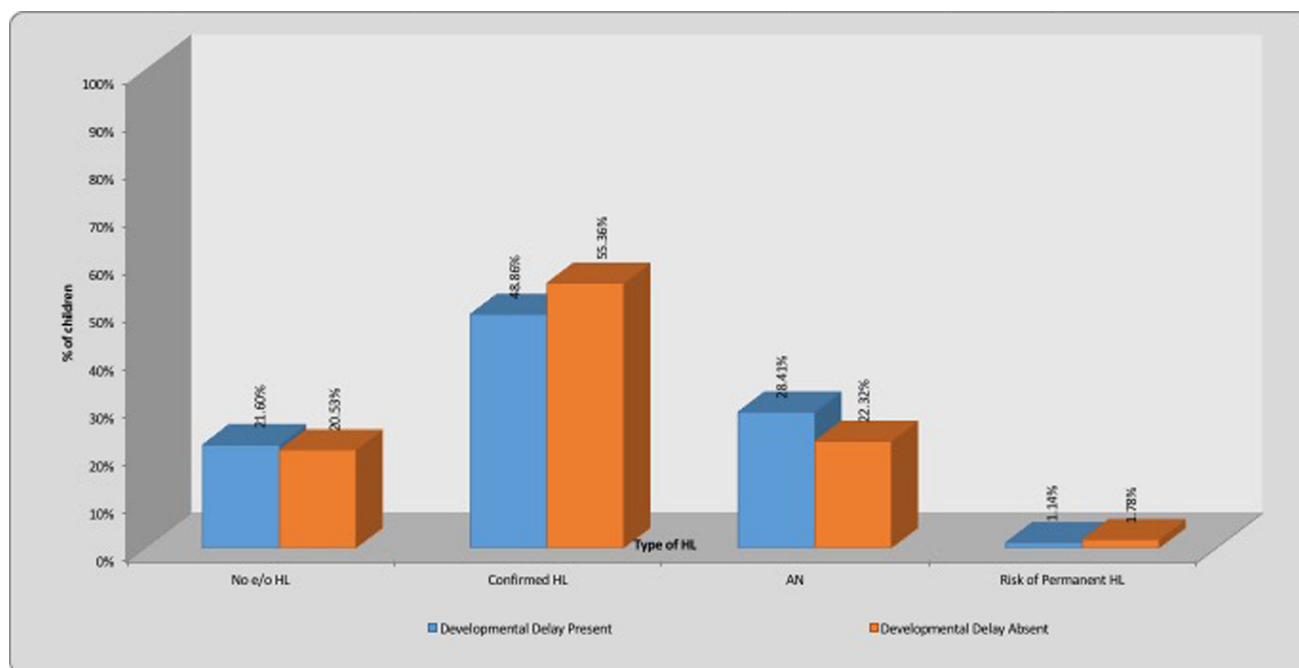


Fig. 3 Association of presence or absence of developmental delay with type of HL

However, the number of ears diagnosed with AN/AD was higher 28.41% in children with developmental delay versus 23.32% children without developmental delay.

Discussion

Otoacoustic emissions (OAEs) and Auditory Brainstem Response (ABR/BERA) have been used in newborn hearing screening programs. The present study was undertaken to study the role of OAE and BERA for identification of pattern of hearing loss, especially the occurrence of suprathreshold disorders (Auditory neuropathy/auditory dys-synchrony (AN/AD) and indirect evidence of Central Auditory Processing disorders) and to correlate them with parental observation of speech or hearing problem. Hearing Loss is an invisible handicap. Hence, many children in our country are not brought for hearing screening by their parents. This is probably first study, which has attempted to correlate presentation of children, that is, hearing and speech problems, with the type of hearing loss, as detected by both OAE and BERA. *The study attempts to highlight the importance of utilizing both these tests, rather only single test for identification of type of hearing loss, especially suprathreshold disorders (Auditory neuropathy/auditory dys-synchrony), and as an indirect evidence of CAPD and mild PHL.*

In the present study, we screened children in age bracket with the youngest child 15 days old while oldest was 5 years old. The largest group i.e. 37 children (37%) were in the age group of 0–1 year. These results were similar to several studies that have reported age of presentation for screening of hearing loss [10–13].

Presentation for Hearing Screening

In the present study, out of 100 children screened, 49 children were brought by parents with complaints related to HL and/or speech defect in their children whereas remaining 51 children who were screened, presented without any complaint related to hearing or speech, but were subjected to screening test for HL on the basis of presence or absence of risk factors. All 49 (49%) children, who were brought by parents, had speech problem and 27% with isolated speech problem. 22% of children (19 children + 3 with suspected HL) in our study, were brought with complaints of HL. Our results were in concordance with other studies where very few children were brought for screening for suspected HL. In the study by Erasmo Gonzalo D. V. Llanes and Charlotte M. Chiong, the majority of the referrals were for speech delay (30.27%), suspected hearing loss (25.29%) or global developmental delay/autism/mental retardation (16.09%),

etc. [10]. In the study by G. Mishra et al., they found that, as the child grows and achieves milestones, the parents, readily identify this handicap [11]. In another study by Thirunavukarasu et al., children who manifested deafness presented late between 9 months–2 years with a similar complaint of delayed speech and language development and those who did not respond to sound [13]. And so, only the suspected deaf and mute children or who are lagging behind their peers are brought to the hospital for screening. Before 6 months of age, identification of deafness is difficult. This fact also supports universal neonatal hearing screening.

Type of Hearing Loss in Children and Association with Developmental Delay

The number of ears diagnosed with AN/AD was slightly higher 28.41% in children with developmental delay versus 23.32% children without developmental delay.

This could be because of common etiologies for developmental delay and AN/AD in the form of birth asphyxia, genetic factors, hyperbilirubinaemia, prenatal infections, etc. [14–16].

Presenting Complaints and Pattern of Hearing Loss as Detected by OAE and BERA

In our study, some form of hearing loss [confirmed hearing loss/auditory neuropathy/auditory dys-synchrony (AN/AD)], was present in 94.74% of ears, on both OAE and BERA testing, in children with profound hearing loss and absent speech. A child who had no hearing loss detected on BERA in both the ears could be a case of *central auditory processing disorder with mild conductive or cochlear hearing loss in one ear, not detected on BERA.*

An important observation in our study was that as many as 72.22% of children had some form of HL (confirmed HL or auditory neuropathy/auditory dys-synchrony), though the children were brought by parents with complaints of speech defect/delay only and were not aware of their hearing impairment. These children with speech defect/delay must always be screened for hearing loss. In other studies, authors have found similar finding [10, 14]. Remaining 27.78% of children having no hearing loss on OAE and BERA, but presenting with speech delay/defect were considered as indirect evidence of CAPD and further investigated with test battery like auditory discrimination tests, auditory temporal processing and patterning tests, dichotic speech tests, monaural low-redundancy speech tests, binaural interaction tests, electroacoustic measures, electrophysiologic measures, for CAPD, wherever feasible.

In 51 children with negative history of speech delay and hearing loss, 42.16% had confirmed hearing loss on OAE

and BERA testing, 30.39% suspected AN/AD (These were the missed children but due to screening they were diagnosed early. These included the children with presence of one or the other risk factors at birth). 27.45% had no hearing loss. These children were kept in follow up and rehabilitated for hearing loss by hearing aids/cochlear implantation. Hence, high risk screening was able to diagnose hearing loss in 72.55% of children, in the absence of any complaints related to hearing or speech. High risk screening is important in developing countries, as a large number of them are found to have some form of hearing loss, without any speech or hearing related problems. Universal screening of all the children by both OAE and BERA would be an ideal situation.

Other studies like, Rughani et al. and Anand et al. have shown that as the number of risk factors increases, the risk of hearing impairment also increases [17, 18].

Another important observation from our study was that 2 children who had OAE refer and BERA pass but presented with speech defect/delay. These are the children with mild permanent hearing loss. Hence, they should be subjected to repeat screening by OAE and BERA and kept in follow up, so that late development of hearing loss may be diagnosed in them [19]. National Center for Hearing Assessment and Management (NCHAM), 2005, had suggested that automated ABR (A-ABR) screening pass after a fail result of an OAE test could lead to sending home some infants with mild permanent hearing loss (PHL) in one or both ears, who would have been otherwise benefitted from early intervention [9]. According to Joint Committee on Infant Hearing (JCIH), all degrees and types of hearing loss in childhood, especially the congenital or developmental causes, of even mild degrees of permanent hearing loss warrant screening and early rehabilitation for better speech outcomes. The current hearing screening protocols and technologies may miss some infants with mild forms of hearing loss [19]. The conventional protocols follow OAE refer and BERA pass as normal hearing (considering low specificity of OAE) and miss children at risk of permanent HL. Such children were few (3children) in our study. Minimum age of screening in our study was 2 weeks, hence every OAE refer was considered significant. The studies which have done screening at birth have high false positivity. However, since in our study, the age group of screening was higher, and two of them presented with speech defects in the absence of any observable HL, OAE refer result, in spite of BERA pass needs attention [8, 9]. More research is needed in this direction based on which, there may be a need to revise the current screening protocols.

A second-stage outpatient screening should be offered to neonates who have OAE fail/BERA pass outcomes. It would help delineate infants, with sensory loss, who would

continue to show OAE fail outcome with normal tympanograms (with a high-frequency 1000-Hz probe tone before 4 months of age), from those with conductive component. In both these situations, follow-up would be indicated. *This discrepancy in OAE and BERA results could be corrected by lowering the intensity of the second step BERA level to 25 dB nHL from the current 35 or 40 dB nHL* [9].

Conclusion

- Both OAE and BERA must be used for identifying the type of hearing loss in children, and may be used to identify suprathreshold disorders [Auditory neuropathy/auditory dys-synchrony (AN/AD)] and indirect evidence of Central Auditory Processing disorders), in case of parental observation of hearing loss and/or speech delay.
- Hearing loss is an invisible handicap. Even in the presence of some form of hearing loss, parents may not be aware of their child's hearing impairment and present, only after it affects child's communication abilities.
- Children with any form of speech defect/delay must be screened for hearing loss. They could have underlying confirmed hearing loss, auditory neuropathy (only retrocochlear hearing loss), or mild hearing loss. Those who are not diagnosed with any form of hearing loss by OAE/BERA, could be suffering from central auditory processing disorder.
- There is a need to re-assess the existing hearing screening protocols by OAE and BERA, as the three step protocol followed in India for hearing screening in children, and also two- step protocol suggested by few, may miss many children with supra-threshold disorders in the form of auditory neuropathy/auditory dys-synchrony and central auditory processing disorders as they may have low or no hearing sensitivity loss and present with speech problems only. Children with CAPD may pass OAE and BERA testing and hence are sent home.
- Early High risk screening can identify hearing loss in children at risk of hearing and/or speech defect.

Limitations

- Neither Universal nor universal high risk screening is being followed in our centre, due to lack of parental motivation for hearing screening in their children, till they find hearing or speech deficits in their children.

- Approximately 50% of the children were screened due to high risk factors, but they also presented late.
- The sample size of children was small to draw conclusion regarding risk of permanent HL in children.

Recommended Future Research

- Better Hearing Screening protocols for large populations at minimum cost and manpower.
- Studies on the role of peripheral/central auditory processing disorders in the causation of speech defects.

Compliance with ethical standards

Human and animal rights All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

References

1. Singh V (2015) Newborn hearing screening: present scenario. *Indian J Community Med* 40:62–65
2. Behrman Richard E, Kliegman Robert M, Jenson Hal B (2004) Hearing loss. *Nelson text book of pediatrics*. Part XXIX, 17th edn. WB Saunders, Philadelphia, pp 2129–2135
3. Homer JJ, Linney S, Strachan DR (2000) Neonatal hearing screening using the auditory brain stem response. *Clin Otolaryngol* 25:66–70
4. Oudesluys-Murphy A, Van Straaten HL, Ens-Dokkum MH, Kauffmane-de Beer MA (2000) Neonatal hearing screening. *Ned Tijdschr Geneesk* 79:69–76
5. Stach BA (2010) Audiologic evaluation of otologic/neurotologic disease. In: Gulya AJ, Minor LB, Poe DS (eds) *Glasscock-Shambaugh surgery of the ear*, 6th edn. Peoples Medical Publishing House USA Shelton, Connecticut, pp 189–221
6. Newborn and infant hearing screening (2009) Current issues and guiding principles for action outcome of a WHO informal consultation held at who headquarters. Geneva, Switzerland
7. Vignesh SS, Jaya V, Sasireka BI, Sarathy K, Vanthana M (2015) Prevalence and referral rates in neonatal hearing screening program using two step hearing screening protocol in Chennai—a prospective study. *Int J Pediatr Otorhinolaryngol* 79:1745–1747
8. Johnson JL, White KR, Widen JE, Gravel JS, James M, Kennalley T et al (2005) A multi-center evaluation of how many infants with permanent hearing loss pass a two stage otoacoustic emissions/automated auditory brainstem response newborn hearing screening protocol. *Pediatrics* 116:663–672
9. White KR, Vohr BR, Meyer S, Widen JE, Johnson JL, Gravel JS et al (2005) A multisite study to examine the efficacy of the otoacoustic emission/automated auditory brainstem response newborn hearing screening protocol: research design and results of the study. *Am J Audiol*. 14:S186–S199
10. Llanes E, Chiong C (2004) Evoked otoacoustic emissions and auditory brainstem responses: concordance in hearing screening among high-risk children. *Acta Otolaryngol* 124:387–390
11. Mishra G, Sharma Y, Mehta K, Patel G (2013) Efficacy of distortion product oto-acoustic emission (OAE)/auditory brainstem evoked response (ABR) protocols in universal neonatal hearing screening and detecting hearing loss in children < 2 years of age. *Indian J Otolaryngol Head Neck Surg* 65:105–110
12. Kumar A, Shah N, Patel KB, Vishwakarma R (2015) Hearing screening in a tertiary care hospital in India. *J Clin Diagn Res* 9:MC01–MC04
13. Thirunavukarasu R, Balasubramaniam GK, Kalyanasundaram RB, Narendran G, Sridhar S (2015) A study of brainstem evoked response audiometry in high-risk infants and children under 10 years of age. *Indian J Otol* 21:134–137
14. Xoinis K, Weirather Y, Mavoori H, Shaha SH, Iwamoto LM (2007) Extremely low birth weight infants are at high risk for auditory neuropathy. *J Perinatol* 27:718–723
15. Adhikari S, Rao KS (2017) Neurodevelopmental outcome of term infants with perinatal asphyxia with hypoxic ischemic encephalopathy stage II. *Brain Dev* 39:107–111
16. Auditory Neuropathy [Internet] (2015). NIDCD. <https://www.nidcd.nih.gov/health/auditory-neuropathy>. Accessed 25 Oct 2017
17. Rughani S, Vyas B, Sinha V, Shah M, Kapileshwarte J, Shah S (2011) Hearing screening in newborns. *World Artic Ear Nose Throat*. 4:1
18. Anand S, Tiwari A, Goyal S (2016) Prospective study for newborn hearing screening—a experience from tertiary care centre in central India. *Pediatr Rev Int J Pediatr Res* 3:668–671
19. American Academy of Pediatrics, Joint Committee on Infant Hearing (2007) Year 2007 position statement: principles and guidelines for early hearing detection and intervention programs. *Pediatrics* 120:898–921

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