

ORIGINAL ARTICLE

FREQUENCY OF CONGENITAL HEARING LOSS IN NEONATES

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Background: Congenital deafness is commonest birth defect and it affects 2–4 neonates among 1000 live births. Detection and intervention especially before 6 months of age prevents severe linguistic, educational and psychosocial repercussions and helps the deaf child in the development of normal speech and language. Children who are identified after 6 months of age experience great difficulties in attaining speech and language. **Methods:** To find out the frequency of hearing loss in neonates, a hospital based observational study was conducted in Combined Military Hospital Abbottabad from June to December 2014. One thousand new-borns selected by consecutive sampling within a specified period of time were subjected to Otoacoustic Emission (OAE) testing. Brain Evoked Response Audiometry (BERA) evaluation was performed in all those who failed OAE testing to confirm the hearing loss. Children born with microtia, meatal stenosis, cleft palate, craniofacial abnormalities and syndromic illnesses were excluded from the study. **Results:** Of 1000 new-borns screened, 465 were males and 535 were females whereas 632 (63.2%) were delivered through C-section and 368 (36.8%) were born via SVD. Four hundred and ninety-one (49%) babies had a positive history of consanguinity among the parents. Out of 1000 infants 13 were having hearing loss which was later on confirmed on BERA evaluation. Among them 7 were males and 6 females, 9 (69%) were born through SVD and 4 (31%) through C-section and 8 (61.5%) new-borns had a positive history of consanguinity among their parents. In all these 13 patients only 2 (15%) patients had profound while the remaining 11 (85%) had moderate to severe hearing loss. **Conclusion:** Frequency of hearing loss in neonates is much higher in our study (13 per 1000) as compared to other parts of the world and demands that more studies should be undertaken on this subject to confirm this.

Keywords: Neonatal screening for hearing loss; Congenital deafness; Hearing loss in neonates; Otoacoustic Emission Scan.

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INTRODUCTION

Inability to hear is the most common disorder in new-borns which is 2–4 per 1000 live births.^{1,2} It can be caused by number of conditions which include high bilirubin levels, use of ototoxic drugs, low APGAR scores, prematurity, low birth weight and TORCH infections during pregnancy.¹ Hearing loss can be inherited or it can be an outcome of an abnormal mutation that occurs during embryonic growth. However, the cause of congenital deafness is never revealed in about 50% of cases.²

Mild to moderate congenital hearing loss in children is commonly detected at the age of 2–3 years² and in such cases, usually the presenting feature is inability to speak or the complains of poor performance in school. On the other hand, children with severe to profound deafness with or without other associated abnormalities of head and neck or syndromic illnesses are detected early.^{3,4} Whatever the cause may be, the hearing loss in children should be detected in the first 6 months of life and intervention must be done during this period.^{4,5} The infants who are treated early have more

chance to develop speech, language and cognitive abilities as compared to those who are intervened after 6 months due to the irreversible changes that occur in their auditory pathways.⁵ Hearing in new-borns can be checked by two different ways: The Otoacoustic Emission (OAE) Scan and the Brain Evoked Response Audiometry (BERA).⁶ These are reliable, non-invasive and automated procedures, which do not need any response from the subject. However, for screening purpose OAE testing is performed because it is easy, cost effective and less time consuming having 99% negative predictive value.⁷ BERA is only utilized to confirm the results OAE scan in positive cases because it is lengthy and time-consuming procedure but more accurate.

The brain's ability to learn speech and language decreases as the child ages, which means that screening for hearing loss is of utmost importance to diagnose children with hearing loss and treat them at early age. There are different methods of intervention that can be used in the children having congenital deafness ranging from hearing aids to cochlear implants with the added support of spoken and sign language.⁸

MATERIAL AND METHODS

A hospital based observational study was conducted in the Out-patient department of Otorhinolaryngology in collaboration with the Department of Gynaecology & Obstetrics, and Paediatrics, Combined Military Hospital Abbottabad with the aim to find out the frequency of congenital hearing loss among the neonates. The duration of the study was 6 months, starting from June–December 2014. One thousand new-borns either delivered through SVD or C-section in CMH Abbottabad, were selected by consecutive sampling. Otoacoustic Emission (OAE) testing was used as a screening method. After performing the otoscopy to rule out the blockage of external auditory canal by amniotic fluid, all the new-borns were subjected to OAE testing. Those patients who passed OAE test were assumed to have normal hearing. Brain Evoked Response Audiometry (BERA) evaluation was performed in all those who failed OAE testing twice to confirm the hearing loss. Children born with microtia, meatal stenosis, cleft palate, craniofacial abnormalities and syndromic illnesses were excluded from the study. The facility of BERA was not present in CMH Abbottabad but it was available in a private sector known as Hearing Clinic with an audiologist who offered his expertise for the completion of this study.

RESULTS

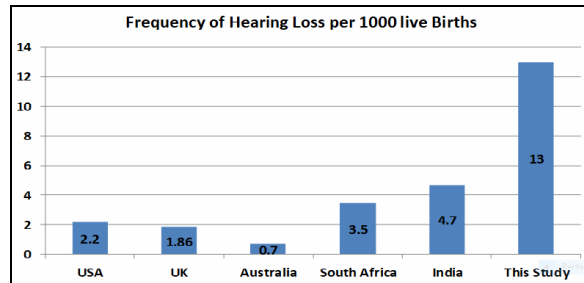
Of 1000 new-borns screened, 465 were males and 535 were females whereas 632 (63.2%) were delivered through C-section and 368 (36.8%) were born via SVD. 491 (49%) babies had a positive history of consanguinity among the parents. Initially 17 cases were found to be positive on OAE scanning; however, on re-scanning after one week, 3 babies passed the test. BERA performed in these 14 patients confirmed bilateral hearing loss in 13 (1.3%) patients and normal hearing in one patient. Out of 13 patients with confirm bilateral hearing loss 7 were males and 6 females, 9 (69%) were born through SVD and 4 (31%) through C-section and 8 (61.5%) new-borns had a positive history of consanguinity among their parents. In all these 13 patients only 2 (15%) patients had profound while the remaining 11(85%) had moderate to severe hearing loss. There was no history of any complication during the pregnancy, prolonged or obstructed labour and all the new-borns cried normally after birth.

Table-1: Showing sex, mode of delivery and consanguinity distribution among 1000 selected new-borns

Sex	
Male	465 (46.5%)
Female	535 (53.5%)
Mode of Delivery	
SVD	362 (36.2%)
C-Section	638 (63.8%)
Consanguinity	
Consanguine	491 (49.1%)
Non-Consanguine	509 (50.9%)

Table-2: Showing frequency of hearing loss, mode of delivery and consanguinity among the 13 affected new-borns

Frequency of Hearing loss	
Hearing Loss	1.3%
Normal Hearing	98.7%%
Mode of delivery in neonates with hearing loss	
SVD	9 (69%)
C-Section	4 (31%)
Consanguinity	
Consanguine	8 (61.5%)
Non-Consanguine	5 (38.5%)



Graph showing frequency congenital hearing loss in different countries of the world

DISCUSSION

Congenital hearing loss is the most common anomaly in neonates. It is more frequent than the conditions that are usually screened at birth, such as hypothyroidism, 25 per 100000 live births, phenylketonuria, 10 per 100000 live births, and galactosaemia, 2 per 100000 live births.² Deafness in neonates is concealed especially if it is not associated with other congenital anomalies of head and neck. The role of early detection and intervention is of great importance for the future development of child.⁸ New-borns having moderate to severe deafness may also suffer problems with speech, psychosocial development and performance in education as compared to other normal hearing children.⁹ In our society, due to lack of awareness and screening programs, parents are usually unable to pick the listening disability of their child before its second birthday or even later when the child fails to develop speech. In our study 11 out of 13 affected new-borns have moderate to severe hearing loss and there is an ample chance that the favourable results can be achieved in the outcome of these children with amplification and appropriate therapy. These children are capable of attaining normal speech, language and developmental milestones. Our study shows 1.3% cases of bilateral congenital hearing loss in our setup which is a huge figure and almost 6–7 times high as compared to some of the studies conducted in different parts of the world.^{2,9,10} Such a high frequency demands that hearing assessment programs should immediately be started in every hospital of the country so that children born with this treatable disability should be managed as early as possible. Although 63.2% of the new-borns in our study delivered through C-section, only 31% were having

hearing loss. On the other hand, 69% of the new-borns having hearing loss delivered through SVD, which shows a positive relation between SVD and congenital hearing loss and may require another study with a larger sample size to confirm this hypothesis. Another significant finding is the ratio of consanguinity among the parents, which is almost 50% in our cases and 61.5% of children with hearing loss are from consanguine couples. Of 1000 new-borns screened by OAE testing only one case of false positive test was discovered which is 7.69% and it is marginally greater than the Colorado study published in 1998. This study does not show any gender significance for congenital hearing loss, this may be because the children born with known risk factors (which are more common in males) were already excluded from the sample.

Our study does not show any case of unilateral deafness, which is relatively a lesser disorder, usually does not hinder the development of speech and language and in most of the cases appears as an incidental diagnosis.^{10,11} However bilateral congenital hearing loss is of more importance because diagnosis after 2 years of age requires heroic efforts for both speech and language development. Even years of untiring efforts may not be sufficient for attaining milestones comparable with normal hearing wards.¹² Screening for congenital hearing loss is obviously a major undertaking and with early intervention, hearing improved children can become normal citizens instead of being burden on the family and society.

The neonates with severe to profound deafness can be benefitted from hearing aids and speech therapy. In addition, surgical options like cochlear implants are now available in our country. Finally, the outcome of this study suggests that the frequency of congenital hearing loss is unfortunately far higher in our region as compared to other parts of the world.^{2,11-14} It demands that more studies should be undertaken on this subject and neonatal hearing screening should be mandatory in every hospital for early detection and intervention, not only, to make our people aware of this common and treatable disability but to improve the life of these effected new-borns in future.

CONCLUSION

Congenital deafness is much more common than expected and future incapacities are too mortifying. It can be detected and managed by neonatal hearing screening which is an attainable realistic goal and can

act as a catalyst in nurturing the lives of the affected children in an unprecedented way.

AUTHORS' CONTRIBUTION

SA: Main Author. Performing OAE scanning, data collecting, analysis and writing. SS: Providing the prenatal and perinatal history, APGAR score and guiding the mothers and sending the neonates for OAE testing. SAM: Helped a lot in getting the BERA testing done from the private sector free of cost. NRA: Provided administrative support and allocation of funds for buying OAE scan. SF: Helped in collection and analysis of data. AR: Helped in making a sound proof room for OAE testing. FB: Counselling of parents of positive Children. ZAN: Counselling of parents of positive children. FEM: Performed BERA of all neonates detected by OAE Scan.

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