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# CLINICAL PROFILE OF PAEDIATRIC COCHLEAR IMPLANT CANDIDATES: A CROSS SECTIONAL STUDY

Otorhinolaryngology

**Dr Adhvaryu Kuthe*** Junior Resident, Department of ENT, GMCH Nagpur *Corresponding Author

**Dr Devendra Mahore** Professor and Head Of Department, Department of ENT, GMCH Nagpur

## ABSTRACT

Hearing Loss is the most common sensory deficit in paediatric group today. With the critical age for language learning period i.e. from birth upto 3 ½ years of age coinciding, the early screening, diagnosis and management for the hearing impaired is a must. Overall 51 children with severe to profound hearing loss were included in the study. Detailed history was noted. Male : Female ratio was 1.217:1. Most common presenting age group was 1-2 years i.e. 31.37%(16). Family History of Hearing loss was found in 7.84 %(4) children and 39.22 %(20) hearing impaired children parents were having consanguineous marriage. Congenital risk factors like pregnancy induced hypertension 7.84%(4), Rh incompatibility 5.88%(3), maternal rubella infection 1.96%(1) and acquired causes like low birth weight 37.25%(19), hyperbilirubinemia 7.84%(4) and meningitis 3.92%(2) with other risk factors were found associated with hearing impairment. Most of the children were full term gestation 68.63%(35) while pre term constituted 29.41%(15). Maximum number of candidates,41.17%(21) attended first consultation at cochlear implant clinic as having hearing loss at the age of 2-3 years. Hearing Aid trial studies showed 50.98%(26) children not benefitting. Radiological abnormalities were seen in 15.69%(8) of children. Cochlear implant was operated on right side in 92.30%(24) and on left side in 7.70%(2) candidates under the benefit of AYJNISHD scheme. This study aims towards identifying the incidence of congenital hearing loss in paediatric age group, the potential risk factors of the congenital sensorineural hearing loss which will prompt early detection with the help of audiological investigations(OAE and BERA). This study also takes note of radiological abnormalities associated with cases of congenital deaf mutism ,if any, before cochlear implant surgery is planned.

## KEYWORDS

Cochlear implant, Risk factor, Hearing aid, Radiology, BERA

## INTRODUCTION

Hearing is one of the major senses and like vision is important for distant communication and for warning about potential risks. Ability to communicate makes man a social animal.

Around 466 million people worldwide have disabling hearing loss and 34 million of these are children. Owing to the fact that hearing is an invisible disability, it may often go undetected until the school going age, especially in children with no additional disabilities. The prevalence of sensorineural hearing loss (SNHL) is 2-3 per 1000 live births in India. Out of 1000 children, 1 child is found to have profound hearing loss at birth or in Pre-lingual age group[1]. Congenital hearing loss is one of the most common congenital anomalies that can be identified early in life due to availability of several sophisticated tests like OAE and BERA.Although neonatal hearing screening tests are important for timely detection and rehabilitation for hearing loss, determining the factors that are responsible for the hearing loss are equally important as it can potentially direct interventions towards prevention of hearing loss.

Brainstem Evoked Response Audiometry (BERA) is one of the popular screening methods for hearing assessment in the neonates and very young children. BERA represents a non-invasive, simple, objective methods for evaluating the function of the auditory apparatus in infants and children. The World Health Organization (WHO) estimates that around 60% childhood hearing loss could be avoided through preventive measures and thereby the child can stand a chance to lead a normal life.

## METHODS

This is a cross sectional study done at a tertiary care centre in Central India from June 2019 December 2021.Patients attending E.N.T. OPD at Tertiary care center between the age 0-5 years brought by parents with severe to profound hearing loss not benefitting with powerful hearing aid trial were included in the study.

**Assessment Is Done Of 3-6 Months For Benefit**

- < 2 years - Based on behavioral observation pre and post hearing aid fitting
- > 2 years – Based on Aided Audiogram pre and post hearing aid fitting

**Assessment After 3-6 Months :**

- Age < 2 years - Failure to show achievement of auditory milestones and behavioral assessment
- Age > 2 years – Child with Aided Audiometry responses below the speech banana curve after powerful binaural hearing aid trial was selected as a candidate for cochlear implant and with Failure to show improvement in single syllable word test of greater than 30%.

The sample size amounted to 51 candidates. A written informed valid consent was obtained from the parents (both mother and father) / guardian using the standard consent form.

A detailed history was obtained from the parents regarding all the trimesters of pregnancy, type of marriage, family history of hearing loss, any infection during pregnancy, use of any ototoxic medication, exposure to radiation, any other illness during pregnancy, time of first consultation at cochlear implant outpatient clinic and hearing aid trial.

Information was obtained regarding the infant/ child- gestational age, birth weight, presence of any complication during labour, admission to NICU for any reason, presence of any post- natal risk factors. Thorough clinical and radiological examination of the child was then performed for anomalies and routine ENT examination. Audiological examination was done by BERA. The presence of risk factor and the subsequent degree of hearing loss was also studied.

## RESULTS

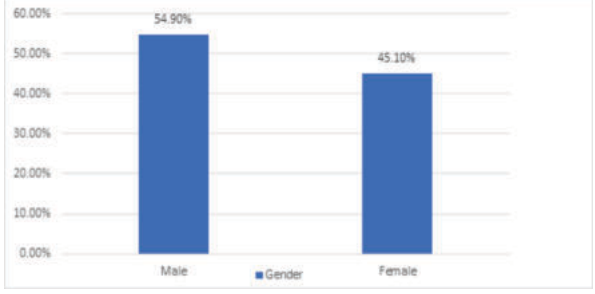


**Fig 1:** Distribution of hearing loss of candidates based on the Gender

In the present study , 54.90 % (28) were boys and 45.10% (23) were girls. There was slight male preponderance in the present study with a Male: Female ratio of 1.217.

**Table I: Distribution Of Candidates Based On Age At Diagnosis**

| Age of Diagnosis (years) | Total no of subjects(n=51) | Percentage (%) |
|---|---|---|
| 0-1 | 10 | 19.60 % |
| 1-2 | 16 | 31.37 % |
| 2-3 | 14 | 27.45 % |
| 3-4 | 9 | 17.64 % |
| 4-5 | 2 | 3.92 % |
| Total | 51 | 100 % |
| Mean(years) | 2.549 | |
| Standard Deviation(years) | 1.119 | |

Out of total 51 subjects, majority 31.37%(16) were in the age group of 1 year - 2 years , age group of 2-3 year constituted 27.45 %(14).

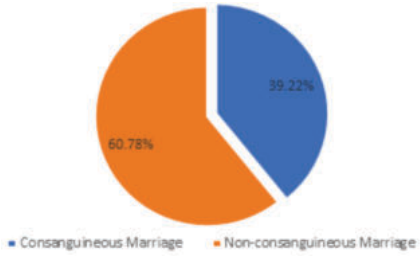


**Fig 2:**Distribution of candidates based on the marriage history of parents

In the present study,Consanguineous marriage was seen in 39.22%(20) children and Non-consanguineous marriage was seen in 60.78%(31) children.

**Distribution of candidates based on the family history of Congenital hearing loss**

In the present study, 7.84 %(4) subjects had family history of congenital hearing loss.

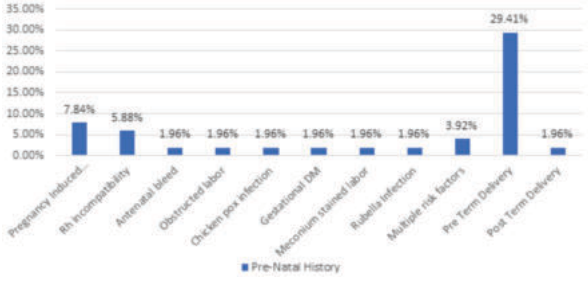


**Fig 3:** Distribution of candidates based on Pre Natal and Gestational Age History(n=51)

- Out of 51 subjects included in the study, 29.41%(15) of these infants and children had presence of one of the prenatal risk factors for congenital hearing impairment. PIH (Pregnancy induced Hypertension and eclampsia) was the most common prenatal risk factor 7.84%(4) present in the study subjects.
- Rh incompatibility was found in 5.88%(3) subjects.
- Obstructed labour was found in 1.96%(1) subject, 1.96%(1) subject had Meconium stained liquor, and 1.96%(1) subject each had rubella infection, chicken pox infection, Ante-natal bleeding and Gestational Diabetes Mellitus as a prenatal risk factor.
- Multiple risk factors were seen in 3.92%(2) children.A Child with prenatal History of PIH and Preterm and another with Obstructed labour and Diabetes Mellitus.
- Pre Term Delivery was seen in 29.41%(15) and Post Term Delivery was seen in 1.92%(1) candidate.

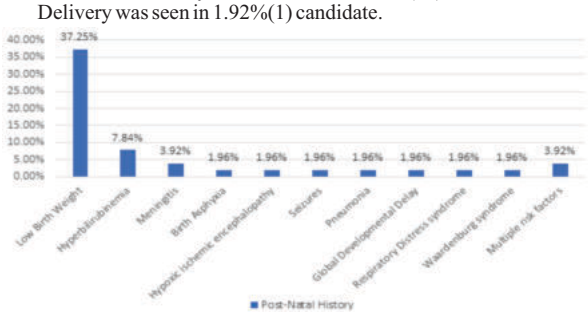


**Fig 4:** Distribution of the candidates based on Post-Natal History

While studying the Post Natal History, 66.67 %(34) of the candidates were having at least any one of the post- natal risk factors known to cause congenital hearing loss. The most common post-natal risk factor that was observed was low birth weight seen in 37.25 %(19). Hyperbilirubinemia was seen in 7.84%(4), 3.92%(2) had meningitis, 1.96%(1) candidate each had pneumonia, Hypoxic ischemic encephalopathy, seizures, Birth asphyxia, Global developmental delay, Respiratory distress syndrome, Waardenburg syndrome.

Multiple post-natal risk factors were seen in 3.92%(2) children.A child with Low Birth weight and Hyperbilirubinemia and another with Low birth weight and meningitis.

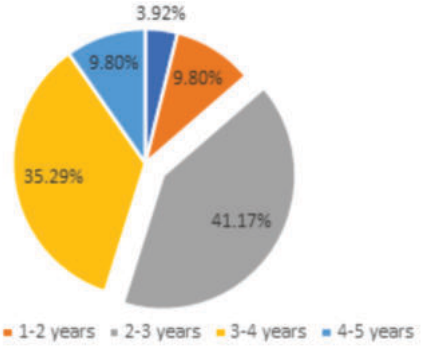


**Fig 5:**Distribution of candidates based on the first consultation to cochlear implant clinic(n=51)

- Maximum number of Candidates attended first consultation at cochlear implant clinic as having hearing loss at the age of 2-3 years constituting 41.17%(21).
- Age group of 0-1 year constituted 3.92%(2).
- In the present study, maximum number of candidates 50.98% (26) did not achieve significant benefit from the Hearing Aid Trial and 27.44%(14) subjects were still under Trial for Hearing Aid.
- Failure to follow up to cochlear implant clinics were seen in 21.5y% (11) subjects.
- After Radiological Assessment, it was found that 8 subjects had Radiological Abnormalities constituting 15.69%(8) of sample size.
- 84.31 %(43) subjects were found to have Normal Radiological scans.
- Out of 8 Radiological abnormalities it was found that 7 subjects had HRCT Temporal Bone Abnormalities constituting 13.72% of sample size.
- Mondini's dysplasia was seen in 3.92%(2), whereas 1.96%(1) subject each had Hypoplastic bilateral cochlea with atresia of apical turn with absence of Semicircular canal, rotated cochlea, High riding jugular bulb, Small internal auditory canal.
- Bilalteral hypoplastic cochlear nerve as detected on MRI was seen in 1.96% (1) candidate.
- In the present study, Cochlear implant candidacy was carefully reviewed and surgery was done in 26 subjects unilaterally as supported by the AYJNISHD scheme.Right side cochlear implant were done in 24 subjects constituting 92.30 % of total subjects while 2 subjects were operated on Left Side constituting 7.70 % of total subjects.

## DISCUSSION

National Hearing Screening program has boosted the detection of hearing impairment at early age and thus empowers parents to make timely choices that will allow their hearing impaired children to be given a good start in life and be fully integrated into the wider community.

In the current study, Hearing loss was found in 51 infants and children. Majority 76.48 % of the children were diagnosed with hearing loss were in the age group of 1-4 years. Thirunavukarasu et al.[2], Desarda et al.[3], Chalak et al.[4], Beigh et al.[5] reported similar findings of majority of hearing loss in the age group of 1-4 years. This high percentage could be because of anxiety among parents regarding the hearing status of their child especially when the child had been exposed to any of the risk factors known to cause hearing loss. This can be attributed to the fact that most of the speech and language development occurs during this period and the hearing loss is identified only when the child presents with delayed speech. BERA can reduce the average age of

diagnosis of significant hearing loss, thus providing an opportunity for early intervention services.

The Male : Female ratio was 1.217:1.Our study findings are in accordance with the different studies conducted by Aiyer et al.(1:1)[6] , Beigh et al.(1.06:1)[5], Jakubikova et al.(0.96:1)[7], Thirunavukarasu et al.(0.91:1)[2]

Family history of congenital hearing loss was found as a risk factor in 7.84%(4) of the subjects similar to Aiyer et al.(7.0 %)[6]

Thirunavukarasu et al.[2] and Zakzouk et al.[8] reported consanguinity (50%) as the most common risk factor in their subjects. This study also found consanguineous marriage as a risk factor in similar percentage (39.21%) of infants and children with hearing loss. Consanguineous marriages can lead to autosomal recessive SNHL. Creating awareness about the association of hearing loss and consanguinity is strongly recommended.

Prematurity and low birth weight were the most common risk factor in studies conducted by Jakubikova et al.[2] as 31%, and Beigh et al.[5] as 20%. In our study, low birth weight was a risk factor in 37.25 %(19) of the subjects with hearing loss.

TORCH infection was reported as a risk factor in 7-10% of congenital hearing loss by Ohl et al.[9], Jakubikova et al.[7] and Watson et al[10]. However, in our study, 1.96 %(1) of subjects with hearing loss had documented TORCH infection post natally.

In the present study, 29.41%(15) preterm had hearing loss. On the other hand, 68.63% (35) full term subjects had hearing loss. Whereas 96%(1) post term subject had hearing loss. Khairy et al.[11] reported hearing loss in 32% of the preterm infants and in 27% of full-term infants studied. However, Ohl et al.[9] reported 60% of the infants having either unilateral or bilateral hearing loss had a history of prematurity. Jakubikova et al.[7] reported prematurity as a risk factor in 31% of the infants with hearing loss.

In our study, most common post-neonatal risk factor encountered was Low Birth weight and Neonatal hyperbilirubinemia. Aiyer et al.[6] also reported hyperbilirubinemia as the most commonly observed post-natal risk factor.Most of the subjects(86.26%) first visited cochlear implant outpatient clinic at 1 year- 4 years of age after being diagnosed with hearing loss. In India, hearing screening facility is mostly available to newborn brought into tertiary hospitals. Some of the key issues in the implementation of the program identified are lack of human resources, inadequate infrastructure, equipment-related shortcomings and low priority for hearing impairment prevention.

The referral to specialized hearing healthcare service should be performed as soon as the hearing loss is detected, in order to prevent social, emotional, and intellectual impairment. Cochlear implant had a positive effect on quality of life of patients and their families .More thoughts need to be given on how to improve hearing health, which involves complex and interdisciplinary planning and technology and how the process of referral and monitoring of patients previously identified as having a disability can be made more efficient. This involves spreading awareness at grass root levels and enhanced communication between centers.

Cochlear implantation is indicated only in those children who do not benefit from a hearing aid. Hence, a thorough hearing aid trial with binaural strong class (i.e. high gain) hearing aids should very essentially be carried out in all children before advocating cochlear implantation.

In the present study, out of 51 total subjects studied with congenital deafness who underwent hearing aid trial, 50.98 %(26) subjects completed the hearing aid trial and were found not to benefit from it and were subjected to cochlear implant surgery.

Out of 51 total subjects studied with congenital deafness, 15.68 % subjects had pre-operative radiological abnormalities. Mondini's Dysplasia 3.92%(2) was the most common finding. Whereas, Otomastoiditis, hypoplastic bilateral cochlea with atresia of apical turn with absence of semicircular canal, Rotated cochlea, High riding jugular bulb, Small internal auditory canal on HRCT scan while bilateral hypoplastic cochlear nerve on MRI constituted other notable findings.

Sangeet Kumar Agarwal et al[12] study findings showed 73% had cochlear anomaly, 87.1% had anomalous vestibule, 56.4% had abnormal vestibular aqueduct, 30.7% had anomalous IAC and 29.4% had abnormal cochleovestibular nerves.

Cochlear implantation surgery in ears with congenital malformation can be troublesome for the surgeon. Major concerns include CSF leakage, the insertion of electrode array, unpredictable distribution of neural tissue in patients with inner-ear anomalies, post-operative meningitis and uncertainty regarding the post-operative outcome. Image studies are, thus, essential before CI surgery. Structural alterations of the cochlea, the middle ear and the mastoid should be identified to help guide the surgical procedure. This approach supports not only selecting appropriate cases for CI surgery , but also preparing surgeons for overcoming abnormalities and avoiding complications that could potentially have a negative effect on the procedure and its results.

## CONCLUSION

In the process of Global epidemiological transition, the economic burden associated with Hearing loss -A Neglected Chronic condition is on the rise, especially in India.

The fact that the age of presentation in our study was majorly more than 1 year makes us ponder why the already placed screening protocol of BERA for those who show Refer/Fail on OAE needs to be popularised at the grassroot levels and so does the counselling regarding referal to higher centres. This will ensure prompt management and therefore can provide an opportunity to initiate an appropriate rehabilitative program at the earliest for the child in the form of hearing aids or Cochlear implantation.

Awareness about the known risk factor will help increase the yield.High-risk neonates have a substantially higher incidence of hearing loss as compared with normal neonates. However, screening of only high-risk neonates is likely to miss some neonates with hearing loss. Hence, screening of normal neonates with no known risk factor should also be considered on clinical grounds.Together we can thus give the child an opportunity to live a near normal life.

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