Prevalence and epidemiology of hearing loss in children attending Cochlea, Pune for hearing and speech

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Abstract

Background:

Cochlea Pune for hearing and Speech is a nongovernmental organization working for rehabilitation of deaf children since 1998. The high burden of deafness in India is largely preventable if appropriate and efficient screening mechanism is in place for entire population.

Objectives:

Present study is an effort to analyse the epidemiology of deafness in paediatric prelingual Indian population attending clinic.

Methods:

A total of 80 subjects screened in the present study following routine medical treatment. Data collected during treatment was analyzed for probable etiology of deafness.

Results:

There is no clear etiology of the deafness for majority of the cases suggesting the need of incorporation of molecular diagnosis and/or more elaborate familial as well as medical history as a part of future screening programs.

Conclusion:

Detection of non-syndromic hearing loss in early childhood is still a challenge in India due to lack of clear policy for post natal deafness screening. Post diagnosis rehabilitation is still a neglected area of deafness management in India. With a decade long experience of deaf screening and management, 'Cochlea Pune for hearing and speech' has all necessary infrastructures to implement 'National Program for prevention and control of deafness' for effective management and rehabilitation of deaf population.

Keywords: Auditory loss, Hearing aid, Non syndromic hearing loss, Cochlea

Introduction

As per the latest report of World Health Organization (WHO), around 7% of Indian population suffer from significant auditory loss, which is the second most cause of disability [1]. Deafness has enormous impact on social, economic and productive life of an individual. Congenital or acquired hearing loss if not detected sufficiently early may result in lifelong deficits in speech and language acquisition, poor academic performance, personal-social maladjustments and emotional difficulties. It has been shown that language acquisition and amelioration of many of the adverse consequences is greatly facilitated if hearing loss is detected as early as six months [2]. Primary health care physicians along with involvement of grassroot workers and community health volunteers has a key role to play for having an effective system of screening in the country of the size and complexity of India.

Considering the enormous social and productive impact of deafness on an individual and thereby on society, government of India has launched a program called 'National Program for Prevention and Control of Deafness' (NPPCD) in 2006. The major components of the program are a) Capacity building and manpower development b) Ear health promotion and prevention c) Early detection of ear problems & management d) Community screening camps e) Rehabilitation and hearing aid provision f) Monitoring and supervision. The efficacy of the program will largely depend on effective implementation with the help of volunteer based nongovernmental organizations (NGO) working in the community health issues.

Cochlea, a nongovernmental organization based in Pune, Maharashtra is committed to reach every deaf child to provide all possible assistance. Cochlea runs a pre-primary school for the deaf children in the age of 3- 6 years and parent-infant program (PIP) for 1-3 years and early detection, early intervention program 0-1 years. Along with that Cochlea conducts certificate teaching courses in collaboration with Rehabilitation council of India (RCI). Present study is aimed at epidemiological analysis of the existing data collected at 'Cochlea' in recent past. The focus of the report is on following key questions –

a) The age of notice and confirmation of deafness and the age of application of hearing aid

b) Types of associated disabilities

c) Etiology of deafness

d) Access to therapy and/or training

Materials and Methods

A total of 80 subjects screened in the present study following routine medical treatment as well as documenting their family and medical history.

As the subjects are minor, parents written consent to use the medical history for preparation of this manuscript has been taken.

Results

A total of 80 patients were assessed in the present study of which 60% were male whereas 40% were female. Patients comprised of both rural (23.5%) as well as urban (76.41%) population. Patients belonging to economically weaker section of society comprise 36.25% of the population.

3.1 Average age of deafness detection and application of hearing aid (HA)

Age of deafness detection and confirmation is vital information for rehabilitation and subsequent choice of therapy. Usually the age of notice and confirmation has discrepancies as detailed patient information is never documented in prescribed format. For present data, patient was asked to mention age of first notice of deafness, age of confirmation of deafness and age of application of hearing aid and speech therapy along with the present age as a part of family history.

Average age of individual child was compared to ascertain the difference between average age of notice & confirmation of deafness and average age of application of hearing aid.

Parents of deaf children approached us for further management of deafness and speech therapy as well as for guidance about further schooling. At the time of investigation average age of the patients in given population was 5.35 years. Average age of notice was found to be 1.70 years. Among the given population 20 percent of children were detected for hearing impairment in their first year of age, implies that 80 percent of cases are not getting opportunity of early detection due to inefficient screening facility. More the children will be detected during screening programs of their first year of the age, language acquisition skills will be far better in such individuals [2].

Average age of confirmation of deafness was found to be 2.02 years. For majority of cases, Brainstem evoke response audiometry (BERA) was performed for confirmation of deafness. Hearing aid was applied at the average age of 2.34 years. None of the patients received hearing aid from governmental schemes.

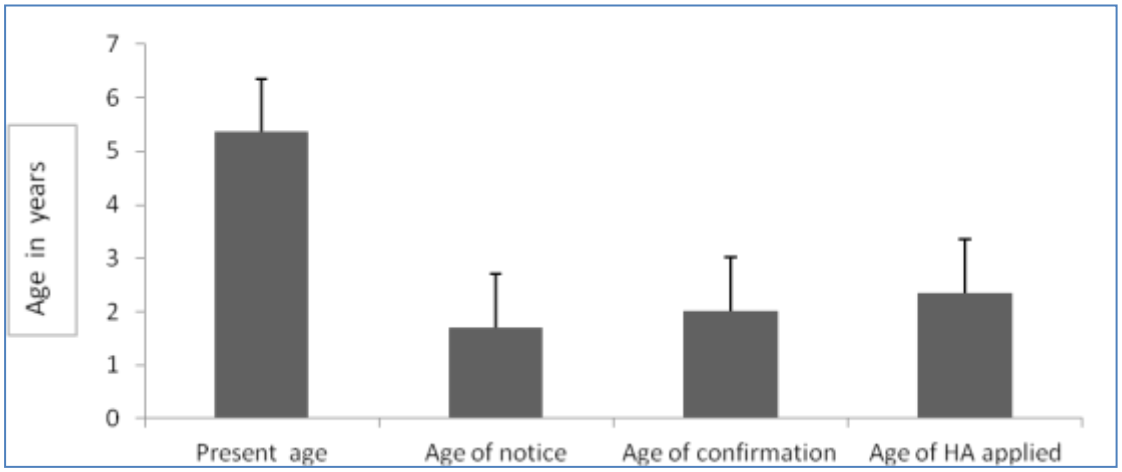

Figure 1: Deafness detection age and application of hearing aid

Average of each type of age is plotted for entire population. Standard deviation was calculated and standard error was plotted for each age type. Average age was found to 5.35 yrs, Average age of notice was 1.70 yrs, Average age of confirmation was 2.02 yrs, and average age of hearing aid applied was 2.34 yrs.

In given population, 50% of the patient could afford to have hearing aid implies that half of the population have no access for hearing aids. Out of 40 cases those opted for hearing aid, 35 undergone some kind of therapy. Out of total 80 cases, 19 are from rural background. Only 8 of rural cases could afford to have hearing aid and among them only three cases have access to therapy. This statistics reveals the status of screening of deafness and availability of trained personnel for deaf population in rural India.

3.2 Reasons of deafness

The primary focus of the study was to find out probable reasons of the deafness in given population with the help of documented family history and diagnostic procedures. At global level, 25% hearing impairment is caused by environmental factors such as prematurity, infections, exposure to ototoxic medications, and trauma. At least 50% of prelingual hearing loss has genetic roots, whereas the etiology remains obscure in the remaining 25% [7].

With given family and medical history, we could correlate the cause of deafness to consanguineous marriages, pregnancy complications or early post natal infections. There were as much as 51.25% patients for whom epidemiology of deafness could not be ascertained. As many as 17.5% cases were of consanguineous marriages and 10% cases were of premature deliveries. There were two cases of family history of deafness in close relatives, and in three cases either or both of the parents were found to be deaf. Infections reported were meningitis, jaundice, hepatitis, typhoid and pneumonia that could be correlated as probable causes of deafness in early childhood in 13.75% of cases.

Only 42% of deaf children have any siblings. Among them only 15% of cases have either elder or younger deaf siblings whereas in 85% cases the siblings were found to be normal.

Pie chart for etiology of deafness in the population

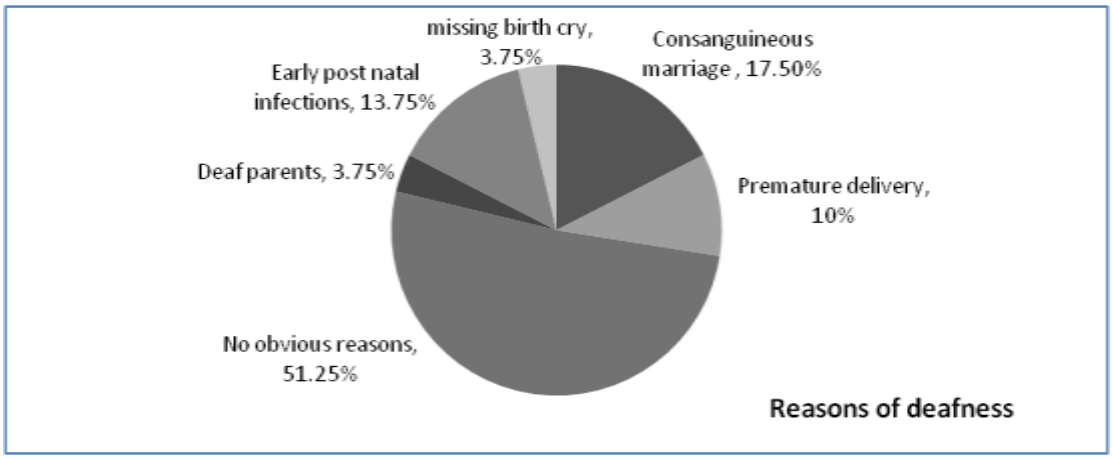

Figure 2: Etiology of deafness in cohort population

The data in given population was collected with medical and family history. 17.5% of cases has marriages in close relation, 10% cases has one or the other type of pregnancy complications, 13.75% cases has history of early infections, 3.75% cases each has birth cry missing and both or either parent found to be deaf, whereas 51.25% cases were without any obvious reason that could be correlated to deafness.

3.3 Associated disabilities with deafness

Deafness though not always but has many times a burden of associated secondary disabilities. In present data we have collected the family history which also mentions any kind of disabilities and/or syndromes. Total 10 cases were reported with various disabilities as tabulated below.

Risk factors associated with a higher incidence of permanent congenital hearing loss include Waardenberg Syndrome, Pendred Syndrome, and Usher Syndrome. Also infections such as cytomegalovirus, rubella, syphilis, herpes simplex and toxoplasmosis, bacterial meningitis are known to be associated in case of syndromic hearing loss [3].

The syndromic disorders are the one where hearing loss is accompanied by involvement of one or several other organ systems and non-syndromic disorders, where the inner ear appears to be the only affected organ.

In present data we have only single case of confirmed syndromic hearing loss. The patient was diagnosed as having hunter syndrome. As the sample size is small, we cannot conclude anything about statistical relevance of syndromic hearing loss in given population.

Table 1: Various disabilities and its corresponding prevalence in a hearing imparied child

Associated disabilities in deaf children	**No. of cases**
Mental retardation	1
Weak motor functions	3
Epilepsy	2
Kidney dysfunction	3
Goitre	1

Discussion

Causes of hearing loss ranges from various environmental factors to the heterogeneous genetic origin adding complexities to identification of etiology. As genetic testing for deafness is not routine procedure, majority of the cases remains without any clear etiology (fig. 2). Genetic form of hearing loss can be syndromic or non-syndromic. In majority of cases hereditary hearing loss is non-syndromic where genetic testing may be useful. Hereditary hearing loss can be inherited as an autosomal dominant, autosomal recessive, X-linked or mitochondrial (maternally inherited) condition. The majority of congenital and pre lingual hearing loss is attributed to autosomal recessive non syndromic hearing loss. It is documented that almost 80% of the childhood

deafness cases are non syndromic [2,4]. In our data barring only one case all others are non syndromic. This may be due to small sample size.

It has been recognized long back that heredity plays role in hearing impairment. The unknown etiology of majority population with prelingual deafness in present study may be attributed to genetic origin. The molecular genetic testing in such cases can be beneficial in improved diagnosis of sensorineural hearing loss in children; it also helps in resolving the diagnostic dilemmas due to variable phenotypes [4,6]. There are no routine diagnostic tests available as the molecular mechanism behind hearing loss is still an emerging field. To date, 57 nonsyndromic deafness genes and more than 1,000 discrete deafness-causing mutations have been described (http://deafnessvariationdatabase.org) [2]. Certain mutations are shown to be prevalent in Indian population [8] but still not have become part of routine diagnosis of deafness.

Present study confirmed the results from earlier reports about need of an effective Information, Education and Communication (IEC) campaign at all levels in the society [5]. Average age of deafness confirmation is found to be 2.02 years (fig. 1) clearly indicates the lapse in effective screening mechanism for detection of deafness within first six months of age. Deafness detected and treated within first year has shown to be most beneficial to formulate better strategies of medication as well as rehabilitation [1].

Only 20% of cases in given population were noticed by parents within one year of age. Deafness detection and treatment in rural India is of concern till date though urban scenario is not far better as far as availability of trained manpower is concerned.

Deaf children and their parents even after getting treatment and hearing aid seemed to be confused about future management of deafness and speech therapy. Average age of patient approaching our clinic was found to be 5.35 years (fig. 1) which is way above their age of confirmation of deafness and application of hearing aid. This clearly suggests that medical and paramedical personnel has major role to play beyond just treatment of deafness and providing the hearing aid.

We also analyzed the patient's history with respect to access to therapy and hearing aid. Only 50% of cases could actually afford to buy hearing aids. None of them obtained the machine with government aid. Only 43% of the deaf children were fortunate to have access to speech therapy at some point of time. When we considered rural and urban cases separately for their access to therapy, only 15% of the rural deaf children had undergone therapy indicating overall apathy towards the deaf population.

There is need of large sample size with more detailed family and medical history to be analyzed for epidemiological analysis of deafness which can be possible under NPPCD. Also universal newborn hearing

screening (UNHS) across the country will help lowering the average age of diagnosis. Of course inclusion of genetic diagnosis may be an added advantage for such screening programs wherever infrastructure and trained man power is available.

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Competing financial interests

The authors declare no competing financial interests.

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Contribution of Authors

PSD has analyzed the data and written the manuscript, AAS has done the audiometric tests. AMW has done the diagnosis and assisted in manuscript writing.

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