Original article

Prevalence and Etiologies of Hearing Loss and Communicative Disorders in Siblings of Children with Deafness

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ABSTRACT

Objectives: The purpose of the study was to estimate the prevalence of hearing impairment and communicative disorder among siblings of deaf children. It also intends to identify etiologies and risk factors of hearing loss to establish the epidemiological data in this population. *Sample:* 622 siblings of the pupils with deafness were subjected to questionnaire and audiometric test. 396 siblings completed the audiometric test. *Design:* The study used the survey research design. Tool: A 24 item questionnaire was prepared to elicit required information to fulfill the need of the research. *Results:* The prevalence of hearing impairment and communication disorder was 26.51% and 16.67% respectively. The study shows that there is huge populations that remain undiagnosed in spite having significant indicators of hearing loss. The study finds positive association between hearing loss and family history of deafness. *Conclusion:* The study underlines the need to increase professional and parental awareness about hearing impairment. Further the study also identified the risk indicators of hearing loss in siblings, this may be used to convince the parents, and hopefully this would improve parental suspicion and increase their demand for audiological testing.

Keywords: Early Identification, Hearing Screening, Communication Disorder, Otitis Media, Family history of deafness

INTRODUCTION

Hearing plays a critical role in the development, comprehension, production, and/or maintenance of speech and/or language (ASHA-2008). Normal hearing provides the primary sensory source for acquisition of language, speech and cognitive skills on which oral communications are instituted. A reduced hearing acuity during infancy and early childhood not only interferes with their psycho-social, linguistic, auditory perceptual and educational development but adversely impacts the family and the society. However, a child's overall future and success can be improved greatly through the early identification of hearing loss, establishment of their causes, and subsequent institution of intervention strategies.

Hearing loss in infants and children may be sensorineural, conductive, or mixed; unilateral or bilateral and symmetrical or asymmetrical. It can also be syndromic (involving other identifiable features) or non-syndromic (isolated hearing loss); congenital or postnatal; prelingual, perilingual or postlingual (i.e. onset before, during, or after speech and language acquisition); and genetic or nongenetic. However, as per WHO (2012) disabling hearing loss refers to hearing loss greater than 40 dB in the better hearing ear in 15 years or older adults and greater than 30 dB in the better hearing ear in 0 to 14 years of children. Hearing loss is an extremely common disorder, with a spectrum of effect ranging from an almost undetectable degree of hearing disability to a profound alteration in the auditory ability

to function in the society. Because its onset is frequently insidious and accompanied by subtle compensatory strategies, hearing loss is usually overlooked by parents, physicians and patients.

BURDEN OF HEARING IMPAIRMNET

Hearing loss is commonest birth defect. The global prevalence of permanent (or sensorineural) hearing loss has been estimated by Mencher (2000) at 1.368 in every 1,000 live births globally. In fact, there is a wide variety estimates of hearing loss worldwide, with ratios ranging between the low of 0.43 per 1,000 (Germany) and the high of 4/1000 (Sierra Leon and Thailand). Fortnum et al (2001) reported adjusted prevalence of Permanent Congenital Hearing Impairment (PCHI) of averages greater than 40 dBHL over 0.5, 1, 2 and 4 kHz in the better ear as 1.07 per 1,000 live births at 3 years and 2.05 per 1,000 live births at more than 9 years of age. Bess et al 1988 reported 11.3% prevalence of minimal hearing loss in school age children. Niskar et al 1998 found 14.9% of children with either low frequency or high frequency hearing loss in a hospital based survey.

According to WHO (2012) estimates 360 million people of world population with hearing loss of 41 dB or more in the better ear, and majority of them said to be living in Asian sub- continents. 32 (9%) million of these are children. Hearing loss is the second most common cause of Years Lived with Disability (YLD) accounting for 4.7% of the total YLD. As per report, deafness is disproportionately high in the Southeast Asia region with a prevalence ranging from 4.6% to 8.8%. Population-based survey (2003) in India using the WHO protocol estimated the prevalence of hearing impairment to be 6.3% or approximately 63 million people suffering from significant auditory loss. The estimated prevalence of adult deafness in India was found to be 7.6% and childhood deafness to be 2% up to the age of 14 years.

The Census of India (2001) reports 1.62 million persons have hearing loss. As per NSSO (2002) 58th round survey found that hearing disability was the second most common cause of disability after locomotor in urban and rural areas. There are 291 persons per 100,000 populations who are suffering from severe to profound hearing loss. Of these, a large percentage is children between the ages of 0 to 14 years. This segment forms 40% of India's population. Based on the reported prevalence, the estimated burden of disease in the school going age group in India is about 26.4 million.

NEED FOR THE STUDY

Several studies indicate variance in the prevalence of newborns with congenital hearing loss. The reason for this big contrast of estimates in different statistical surveys in India are due different settings, population studied and criteria for screening used, and no wonder that available data and enormity of problem is grossly underestimated. Surprisingly, prevalence of hearing impairment as per Planning Commission, Rehabilitation Council and PWD Act (1995) of India are still awaited. Most children with congenital hearing loss have hearing impairment at birth and are potentially identifiable by newborn and infant hearing screening. However, some congenital hearing loss may not become evident until later in childhood.

However, unlike the developed nations, at present, no formal procedures exist to screen children for hearing impairment in India. Since, case histories often reveal a positive family history of communication disorders. Between 28% and 60% of children with a speech and language deficit have a sibling and/or parent who are also affected. Therefore, epidemiological studies estimating the prevalence of permanent hearing loss in children at various ages are of paramount importance to set priorities for prevention and treatment (Parving, 1999). Given the high prevalence of this condition in babies, infants, and young children (Northern & Downs, 2002), and it's devastating consequences resulting in life-long disability if left untreated (Northern & Hayes, 1994), permanent hearing loss in children must be acted upon diligently and as early as possible in life (Yoshinaga-Itano, Sedey, Coulter, & Mehl, 1998).

Moreover, JCIH (1972) in its supplementary statement included a positive family history of childhood deafness as a potential risk criterion for hearing screening which remain unaltered in recent position statement 2007. Furthermore, Optimal Surveillance in the Medical Home (JCIH, 2007) recommended that if hearing loss is diagnosed in infant, siblings of infant should be referred for audiological evaluation by 24-30 months of age.

Additionally, though hearing health care professionals are loud enough at every platform and unequivocally agree to the need of hearing screening programs and continue to blame our co professionals of being un co-operative and insensitive to the cause of hearing screening in developing and under developed countries. But, hearing care professionals especially audiologist and otologist have not yet paid any heed to the recommendation to screen the siblings of hearing impaired child. This risk indicator for hearing loss has been put forth since many decades by professional bodies.

Within these contexts, the present study was conceptualized to assess the prevalence of hearing loss in siblings of children with hearing impairment as there is no study has been yet reported considering the recommendation of JCIH 2007 and as per WHO criteria of disabling hearing loss in this population to best of our knowledge. Such study may be able to sensitize hearing health care providers like otologist, audiologist, special educators and pediatrician etc to conduct hearing screening on siblings of deaf child or at least vigil the parents about hearing loss in other children.

OBJECTIVES

To determine the prevalence of hearing loss and communicative disorder in siblings of children with hearing impairment.

To identify risk factors in this population so as to establish the epidemiological data that can be used to sensitize hearing health care professionals to initiate screening in the siblings of children with hearing impairment.

METHOD

Sample:

The siblings of the pupils with known hearing impairment visiting to our clinic or attending special schools in and around the Mumbai city were invited to participate in the study. 750 siblings were invited to participate in this study, of which 622 accepted the Invitation.

Design:

The study used survey research design to collect the data to fulfill the need of the research undertaken. The necessary consent was obtained from the subject to participate voluntarily.

Tool:

A 24 item questionnaire was prepared to elicit their demographics, clinical symptoms and the main risk factors for hearing loss and communicative disorders. The demographic data includes age, sex, number and birth order of siblings, number and age of siblings known to be hearing impaired, previous hearing evaluation and audiogram. The clinical symptoms and risk factors for hearing impairment and communicative disorder included family history of hearing impairment, consanguinity of parental relationship, birth weight (<1500g) and gestational age at birth (<33 weeks), complications during the pregnancy, and history of ear surgery, infections, trauma, previous ear infections, history of jaundice requiring intervention, meningitis, use of ototoxic drugs and exposure to noise.

The audiometric test was performed using calibrated pure tone audiometer in standard test environment. Subject who demonstrated communication disorders on questionnaire were subjected to speech and language evaluation.

Procedures:

The Questionnaire was presented to parents or siblings of children with hearing impairment, to be answered by the parent/guardian to elicit the responses. Following

the questionnaire, the siblings old enough to complete the test satisfactorily were subjected to audiometric evaluations. Each audiological assessment involved otoscopy, Rinne and Weber test (performed with 512 Hz Tuning fork) and pure tone audiometry at octave frequencies from 500 Hz to 4 kHz. Pure tone thresholds were acquired from 500 to 4000 Hz via air and bone conduction using Hughson Westlake procedure.

According to the findings of these tests, a participant was considered to have normal hearing, conductive, sensorineural or mixed hearing loss. Any individual found to have an average threshold of >25dBHL, over 500 to 4000Hz frequencies in their better ear was classified as hearing impaired. Hearing loss was further classified into mild (26-40 dB), moderate (41-55 dB), moderately severe (56-70 dB), severe (71-90 dB) and profound (>91 dB), as per WHO classification. Only siblings who underwent audiometry were included for the analysis.

Statistical Analysis :

Descriptive analysis of demographics was done for all subjects and prevalence of degree and types of hearing loss was calculated. Comparative analysis between siblings with normal hearing and hearing impairment was done for risk factors. Pearson chi-squared was used at a significance level of 0.05.

RESULTS

Demographic data

Total 622 siblings responded to the questionnaire, Table 1 indicates means, standard deviation median and range (ages in years) of male and female siblings of children with hearing impairment. 519 siblings attended the audiometric session. 396 (76%) siblings completed pure tone audiometry. 123 (24%) children were either underage or unable to complete pure tone audiometry satisfactorily. Of those tested, 201 (51%) were male with mean age of 8.9 years and 195 (49%) female with mean age of 9.6 years.

Table:1

Showing means, standard deviation and median (ages in years) of male and female siblings of children with hearing impairment

Population	Numbers	Mean	S.D.	Median	Age Range
Male	331	8.9	4.4	7.8	4 - 17
Female	299	9.6	4.8	7.1	4 - 15
overall	622	9.3	4.6	9.4	4-17

Table 2

Showing percentage of gender distribution of hearing loss found in siblings of children with hearing impairment

Population	Total number of siblings	Number ofsiblings with hearing loss	p- value
Male	201	64 (32%)	
Female	195	61 (31%)	0.74
overall	396	105	

Prevalence of Hearing Impairment and Communicative Disorder

All individuals had at least one sibling known to be hearing impaired. Individuals had a family composition of mean 2.1 male siblings and 1.8 female siblings. Table 2 Showing percentages and gender distribution of hearing loss found in siblings of children with hearing impairment

A total 105 (26.51%) out of 396 individuals were found to

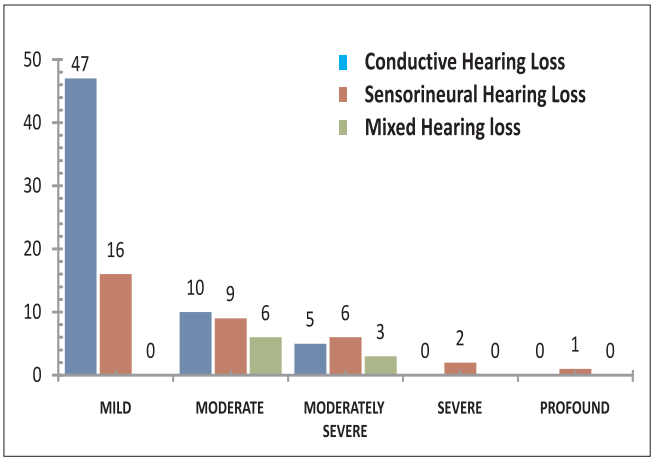

Figure 1. Showing the break up for type and degree of hearing loss in siblings

have a hearing loss of > 25 dB, in their better ear. Distribution of hearing loss between the sexes was insignificant at p=0.74. 64 (32%) males and 61(31%) females out of 105 suffered with hearing loss. Among them 61 (58.09%) had conductive, 34 (32.38%) had sensorineural and 9 (8.57%) had mixed hearing losses. Degree of hearing loss seen from mild 63 (60%), moderate 25 (23.81%), moderately severe 14 (13.33%), 2 (1.90%) severe and 1(0.95%) profound loss.

Prevalence of Communication Disorder

The study records communication disorders in 70 (17.67%) out of 396 siblings with normal hearing and hearing impairment. Table-3, showing the frequency and distribution of communication disorders in siblings having normal hearing and hearing impairment. Voice problems, misarticulations, Stuttering and followed by delayed speech and language, was prevalent at the rate of 31.42%, 24.28%, 21.42 % and 20% respectively.

Table-3

Showing the frequency and distribution of communication disorders in siblings having normal hearing and hearing impairment

Risk factors	Normal Hearing n-291	Hearing Impairment n-105	*p*- Value at 0.05
Family history of deafness	40 (13.7%)	27 (25.7%)	**0.023**
Consanguinity	98 (33.6%)	34 (32.3%)	0.91
birth weight <1500 grams)	63 (21.6%)	21 (20.0%)	0.58
Prematurity < 33 weeks	54 (18.5%)	21 (20.0%)	0.78
Complications during pregnancy	15 (5.15%)	9 (3.09%)	0.743
History of ear infection	29 (9.96%)	39 (37.1%)	**0.002**
Jaundice	18 (6.18%)	11(8.57)	0.56
Meningitis	5 (1.71%)	3 (2.80%)	0.0521
Antibiotic consumption	9 (9.96%)	7(37.1%)	**0.012**
Noise exposure	19 (6.52%)	17 (16.2%)	**0.034**

Table-4

Comparison of risk factors of hearing impairment in siblings of children with hearing impairment

Risk factors	Normal Hearing n-291	Hearing Impairment n-105	*p*- Value at 0.05
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Risk Factors for Hearing Impairment

The risk factor causing hearing loss in the sibling of children with hearing impairment was explored. The risk factors that resulted in hearing loss in the siblings of hearing impairment (depicted in Table-4) statistically significant at *p*- value 0.05.

The most prevalent causes of conductive hearing impairment were found to be chronic supportive otitis media and primary reasons for sensorineural hearing loss was consumption of antibiotic, noise exposure followed by family history of deafness.

DISCUSSION

We screened and evaluated hearing impairment and communication disorders in population that falls in the high risk registry and in which the concept of hearing impairment should be apparent to the professionals and parents. Despite this awareness, the prevalence of undiagnosed hearing impairment amongst this population is unacceptably higher than general populations. The prevalence of hearing impairment and communication disorder in studied population found to be 26.51% and 16.67% respectively. Which is higher than the figures quoted by NSSO 2002 in 58th round as 15% and 10% of hearing and speech problem respectively in general population.

Bess et al 1988 reported 11.3% prevalence of minimal hearing loss in school age children. Niskar et al 1998 found 14.9% of children with either low frequency or high frequency hearing loss in a hospital based survey. The equivalent figure can be estimated from our study, 42 (10.60%) of the 396 were found to have a hearing loss of > 40 dB (Figure1). Most significant prevalence rate of hearing loss to our study has been reported by Abdel Rahman et al (2007). They reported 22.2% of hearing loss in secondary school going children in Ismailia, Egypt.

Though, Fortnum et al (2001) reported that the prevalence of sensorineural hearing loss using criteria of > 40 dB of 1.3 per 1000 live births. Data from Sweden indicated a prevalence of 2.6 per 1000, with 30 dB as the cut off and from Germany the prevalence rate is 4.3 per 1000. But these three reports are from countries where antenatal cares are better and identification of hearing loss is actively sought at an early age. Whereas, countries with fewer resources available for the detection of hearing loss have reported higher prevalence rate like Giles et al (1991) found 120 per 1000 Maori school children to have a hearing loss > 20 dB which is more closer to our findings. WHO (2012) estimates 360 million people worldwide with hearing loss of > 41 dB in the better ear and majority of them live in Asian and African countries.

In spite, high prevalence of hearing impairment among the siblings of hearing impaired children, very small number of children 14.2%, had previous audiogram as reported by parents. Ironically, the identification age of hearing loss in the second child was greater. Though, data was not categorized into mean ages of identification and degrees of hearing loss. However, the mean age of diagnosis of hearing loss by professionals in current study was 8.1 years.

The study noted various reasons of high mean age of diagnosis. On inquiry, the parents revealed that non availability of finances for treatment of second child, improper guidance and testing by professionals, lack of transportation facilities and other domestic conditions were major reasons. With such a high prevalence, mean age of undiagnosed hearing loss and untreated hearing loss indicates that we must strive to promote professional and parental awareness for assessment of hearing impairment in siblings of deaf children and to encourage not only screening programs specifically aimed for siblings of hearing impaired children but also community antenatal healthcare for expecting mothers and hearing screening programs in general population.

The study records only 7 parents with hearing impairment. However, a positive family history was noted in 67 of the screened population. 40 (13.7%) and 27 (25.7%) children found to be having normal hearing and hearing impairment respectively. A genetic component

seems likely, given the statistically significant *p*-value of 0.023 in siblings having hearing loss with family histories. However, we are limited in commenting on this issue since the cause of each individual's hearing loss was not established.

There was a high rate of consanguineous marriages in the population, but no association was found between consanguinity and hearing impairment. However, Elahi et al (1998) stated that in children with a severe hearing loss, 70% could be due to consanguineous marriages. Snashall (1998) reported that consanguineous marriages may result in prevalence of hearing loss of 12 per 1000 population. Further he quoted that factors like a positive family history, congenital abnormalities of the head and neck, prenatal and perinatal infections, prematurity, low birth weight, anoxia and hyperbilirubinaemia may increase the prevalence of hearing loss in affected children.

Hence, it can be concluded that the high prevalence of hearing loss in current study may also be due to all above compounding factors as consanguineous marriages, family history, prematurity and low birth weight were noted in studied population. The risk variable also indicated that children who were exposed to noise had hearing loss at statistically significant value of 0.034. This is possible as India is one the loudest country in the world; none of its social, religious and cultural activities are without the deafening loudness.

There were about 86 children with a mild-moderately-severe hearing loss primarily due to conductive pathology which is almost entirely environmental in etiology. ICMR (1983) reported conductive hearing loss as high as 40% in rural India. The conductive hearing loss is significant at *p*-value of 0.002 in children with positive histories of ear infection. It is likely that majority of these hearing losses would have been due to otitis media, as evidenced by the link between a histories of ear infections and hearing status. It is important to note that all these children are attending regular school, but none of the schools have ever reported and noticed hearing difficulty in these children, rather school labeled them as slow learner and advised private/extra coaching for children. This warrants that hearing health awareness campaign should be done among school administrators and teachers. The school should offer hearing screening for all pupils or at least to children who are having risk indicators and poor academic performance.

CONCLUSION

This study has revealed an unacceptable level of hearing impairment and communicative disorder in siblings of children who have deafness, and whose treating professional and parents are already aware of the impact of hearing loss in children. In spite of awareness neither hearing health care professionals nor parents and school sought any assessment for them. Hence some form of screening and testing procedure must be implemented to curb this high rate of undiagnosed hearing loss. Though, we are aware that the screening for siblings of children with hearing impairment has been advocated by the JCIH 2007 and other organizations but still we fail to screen these children or motivate parents to seek our services for their undiagnosed children may be because of our costly services and non-availability professionals and facilities. Therefore, we must offer services at reasonable cost and at reachable place.

The study also identified the risk indicators of hearing loss in siblings; this may be used to convince and promote health of the child by providing counseling regarding medically and surgically correctable causes of conductive hearing loss. Hopefully this would improve parental suspicion and increase their demand for audiological testing. Considering the significant occurrences of undetected hearing loss and communicative disorder in siblings of hearing impaired child warrants immediate attention towards organized early identification and intervention program through screening procedures. However, till such times arrive in our country, we would like to put forth following recommendation for immediate considerations to all concern.

1. In the absence of any screening program in country like ours, we would advocate that National Programme for Prevention and Control of Deafness in collaboration with Sarva Siksha Abhyian should conduct hearing screening for this group of children.
2. Further an active campaign is needed to heighten the awareness of hearing impairment in siblings of hearing impaired children among parents, medical professionals, educators and general public.
3. Awareness program should be developed for professionals about medically and surgically non treatable hearing losses and necessary training of the identification and treatment of hearing loss.
4. Training program to identify developmental disabilities and available intervention resources in the form educational material should be made available to the educators at school.
5. There is need for more qualified personnel to treat ear pathologies and auditory deficits and provide speech and language training so that services can be made available in the vicinity.
6. Fund raising is required for poor and needy people. If possible subsidized or free of cost services should be provided even to lesser degree of hearing loss conditions that is not available at this point of time.
7. Last but not the least, Audiologist, as hearing health care professional if we find any child with hearing impairment we should offer his/her other siblings for some form of hearing screening free of cost as a social responsibility to promote hearing health in them.

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