Prevalence of Hearing impairment in Sibilings of Deaf Children

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Abstract

Objective: To assess the prevalence of hearing impairment in siblings of children with profound to total hearing impairment.
Method: Two hundred and twenty siblings of children at a deaf school in Karachi completed the screening program. This consisted of a questionnaire and pure tone audiometry.
Results: One out of three (73 siblings) had a threshold of 25 dB or greater in their better ear. In a population, already aware of hearing loss, we note that only fifteen siblings had a previous audiogram.
Although a positive family history is associated with hearing impairment in the siblings, no increase due to consanguineous marriages was noted.
Conclusion: This assessment has underlined the need to increase public awareness of hearing impairment. It has also exp.osed the lack of it in families exposed to the condition (JPMA 52:73,2002).

Introduction

Unlike the developed world, at present, no formal procedures exist to screen children for hearing impairment in Pakistan. Many studies[1-4] have investigated the prevalence of childhood hearing impairment, based on a variety of criteria, in settings throughout the world. For our purposes, the most pertinent data is probably that published by Elahi et al[5] and Zakzouk et al[6], who used the same criteria to define 'hearing loss' as our study. Elahi et al[5] found the prevalence of all hearing loss in rural Pakistan to be 7.9%, in a general population setting. Interestingly, Z:ikzouk et al[6] faund that children, with hearing impaired siblings, were more likely to have an undiagnosed hearing loss themselves. They quote the prevalence of hearing impairment among those children, with hearing-impaired siblings, to be 24.8%, compared to 7.7% among all children screened, in Saudi Arabia.
The Ear Foundation Pakistan (EFP), a field charity for the hearing impaired, chose to screen the siblings of hearing-impaired children, attending the ABSA (Anjumane-Bahbood-e-Samat-e-Atfal) School, Karachi. Having 473 pupils, the charity-funded school caters for severely and profoundly hearing impaired children.

Subjects and Methods

The siblings of the pupils, attending the ABSA School for hearing impaired children, Karachi, were invited to take part in audiometric screening. A questionnaire was presentcd to each sibling, to be answered by the parent/guardian. Questions were intended to elicit the following aspects of their histories: age and sex, birth order in the family, birth weight (<1 500g) and gestational age at birth (<33 weeks) complications during the pregnancy, number and age of siblings, known to be hearing impaired, family history of hearing impairment, consanguinity of parental relationship, whether a previous audiogram had been performed, previous ear infections, history ofjaundice, requiring intervention and, history of meningitis.

Following the questionnaire, pure tone audiometry was performed on siblings old enough to complete the test satisfactorily. Any individual found to have a mean threshold of ~25 dB, over 0.5k, 1k and 2k frequencies in their better ear was classified as hearing impaired.

Hearing loss was further classified into mild (25-44 dB), moderate (45-64 dB), severe (65-94 dB) and profound (>95 dB), as described by Davis7. Only siblings who underwent audiometry have been included in the following results. Analysis was performed using chi-squared statistical evaluation.

Results

Two hundred and forty-eight siblings attended the screening session. Two hundred and twenty underwent pure tone audiometry. 28 (12.7%) children being underage to complete this test satisfactorily. Of those screened, 105 (48%) were male and 11 5 (52%) female. Ages ranged.from 4 to 25 years (mean age of 11) in males and from 4 to 23 years (mean age of 11) in females. Individuals had a family composition of mean 2.4 male siblings and 2.2 female siblings. All individuals had at least one sibling known to be hearing impaired, attending the school. Each had a mean 0.9 male hearing impaired siblings and a mean 0 5 femaIe hearing impaired siblings.

Seventy-three (33.2%) individuals were found to have a hearing loss of >.25 dB, in their better ear.

Distribution between the sexes was insignificant (p=0.74); 36 (34.2%) males and 3,7 (32%) females. The mean age of these children was 11.1 years.

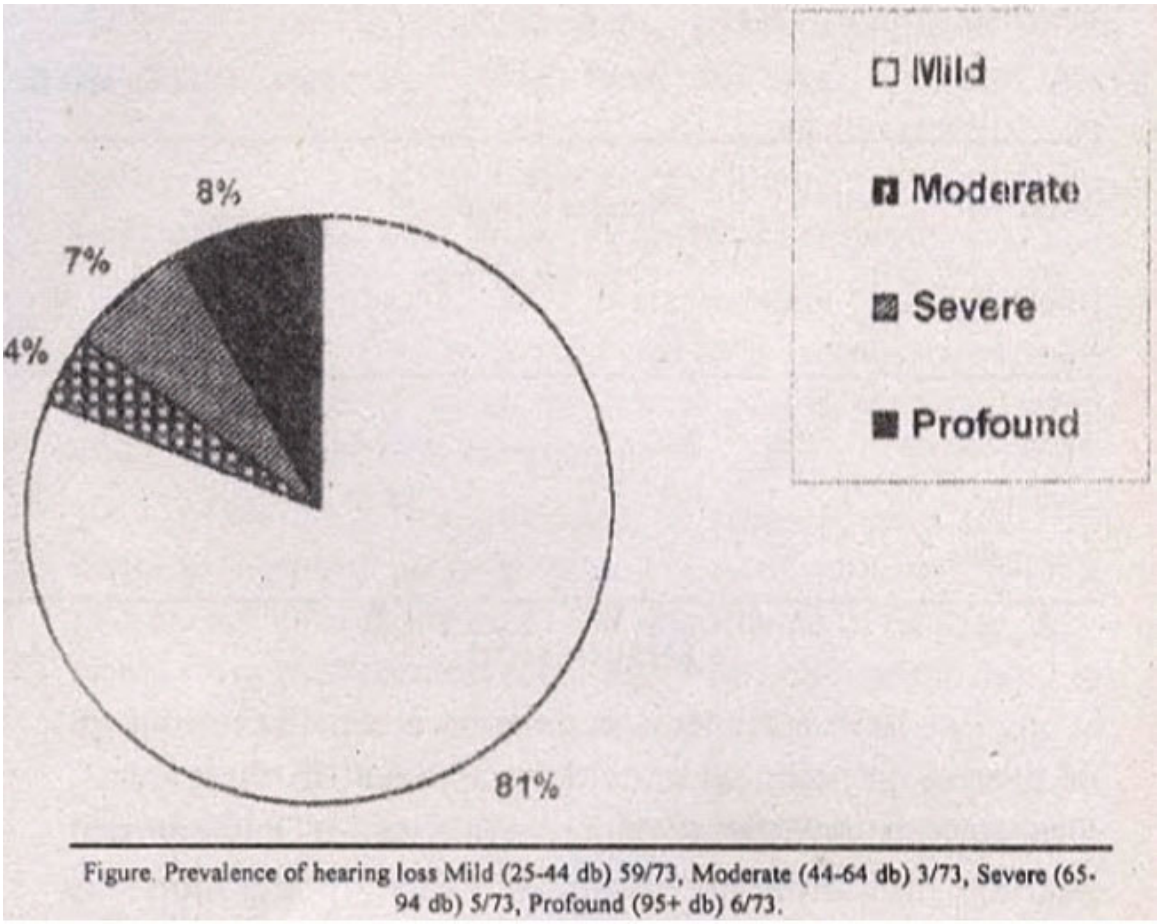

Figure. Prevalence of hearing loss Mild (25-44 db) 59/73, Moderate (44-64 db) 3/73, Severe (65-94 db) 5/73, Profound (95+ db) 6/73.

Figure shows the breakdown of hearing loss into the categories of mild, moderate, severe and profound loss.

There was no association between a child's birth order and their hearing status. Sixty-nine (31.5%) of the siblings were born at home. None were born prematurely. Eleven were born with birth weight <1500 grams. Two of these eleven were identified as being hearing impaired (p=O.58). Complications during pregnancy, other than hypertension (7.2%) were rare.

In all those screened, 140 (63.6%) of parental relationships were consanguineous. Consanguinity had no effect on hearing status in hearing impaired population; 63% (46/73) of hearing impaired individuals compared to 64% (94/147) of normal hearing individuals (p0.89).

Parental deathess was positively stated in only 2 fathers and I mother (different families). The children of the hearing impaired fathers showed no hearing loss, whereas the child of the mother was noted to have a mild hearing loss.

A positive family history of deafness, in relatives other than parents and siblings, was elicited in 18.2% (40/220). A positive family history did appear to be linked to hearing impairment in the screened siblings. Of the siblings found to have a hearing loss, 26% (19/73) had a family history, whereas 14.3% (2 1/147) of normal hearing siblings had a family history of hearing impairment (p=O.O13).

Previous audiograms had been performed in only 15 of the 220 screened children (6.8%).

Of those identified as being hearing impaired 137% (10/73) and 3.4% (5/147) of those with normal hearing had a previous audiogram (p=.004).
The results for positive histories of risk factors for hearing impairment; ear infections, jaundice and meningitis, are displayed in Table.

Table. Risk factors for hearing impairment.

History of	Number of impaired hearing siblings		Number of normal hearing siblings		Statistical difference X^2
	No.	%	No.	%	
Ear infections	20/73	27.4	16/147	10.9	p= 0.002
Jaundice	6/73	8.2	9/147	6.1	p= 0.56
Meningitis	1/73	1.4	7/147	4.8	p= 0.21

employed to define hearing loss, it is clear that the prevalence in our population is far higher than those quoted by other population studies.
Our findings closely mirror those of Zakzoük6 They also found the prevalence of hearing impairment, among children whose siblings have a hearing loss (24.8%), to be far higher than in the general population (7,7%).

Discussion

We have screened a population in which the concept of hearing impairment should be apparent to the parents. Despite this awareness, the prevalence of undiagnosed hearing impairment amongst this population is unacceptably higher than other estimates, performed on general populations. The point prevalence of hearing impairment in our screened population was 33.2%. The figure quoted by Elahi et al[5], having screened a population of children in rural Pakistan, was 7.9%. Other studies publishing data on the prevalence of childhood hearing loss have used different criteria to define hearing loss[1,3,4].
Fortnum et al[1] found the prevalence to be 1.3 per 1000 livebirths. in a representative UK population. Using their criteria of> 40 dB, they found most hearing loss to be sensorineural,. The equivalent figure from, our study is 192 per 1000 children screened, since 14 of the 73 were found to have a hearing loss of ?. 40 dB. Data from Sweden[3] indicate a prevalence of 2.6 per 1000, with ? 30 dB as the cut off and from Germany[4] the figure is 4.3 per 1000.
These three studies[1,3,4] have been conducted in countries where identification of hearing loss is actively sought at an early age. With established screening programs in the UK, it is interesting to note that Asian children were found to have an increased risk of sensorineural hearing loss, over non-Asian children[8]. More pertinent to our study are the figures for countries with fewer resources available for the detection of hearing loss. 0 lIes et al[2] found 120 per 1000 Maori school children to have a hearing loss ?20 dB. With the appropriate corrections made to the criteria, Of the 541 invited for screening, nearly

half attended, it is likely that there was a degree of bias in those attending, with many refusing, assuming their hearing to be normal. Unavailability of transport and parents' domestic situations were other reasons for non-response. Even taking this into account the prevalence is high.

Coupled with the high prevalence of hearing impairment the number of children who had a previous audiogram (6.8%) is small, It would be fair to assume that having experienced the effects of hearing loss in one of their children, parents might be eager to rule out a similar impairment in other children. Although the vast majority of the children identified suffered with mild to moderate hearing loss, but five had a severe and six profound hearing inpairment. In a population of hearing impaired children in Germany[9] strong correlation was found between the degree of hearing loss and age at which this was diagnosed. On an an average children with a profound hearing loss were diagnosed at 1.9 years, those with a severe loss at 2.5 years, with a moderate loss at 4.4 years and with a mild loss at 6.2 years.

It is stated that the mean age at diagnosis remains very high. Although not categorized into degrees of hearing loss, the mean age in those children found to be hearing impaired in this study was 11.1 years. A lot more therefore needs to be done to promote parental awareness for assessment of hearing impairment in siblings of their deaf children and to encourage not only community screening programs, but also programs aimed specifically at siblings of hearing impaired children.

Parental suspicion has been shown to be a poor indicator of childhood hearing impairment. Watkin et al.[10]

Lound those parents only suspected a hearing impairment in 44%~ of children with severe or profound hearing loss and even lower for those with a mild or moderate hearing loss. With such a high prevalence and high mean age of undiagnosed hearing loss in our population, it would appear that parental suspicion was lower still in this group.

One plausible explanation for these findings is that parental perceptions of hearing are influenced by their experiences with their children, or "close relatives", who are known to be hearing impaired. For instance, compared to a severely hearing impaired child, a sibling with mild to moderate hearing loss may be perceived as 'normal' despite having a condition which could lead to poor speech and educational development.

Only three parents were identified as hearing impaired in this population. However, a positive family history was noted in 18.2% of the screened population and 26.0% in those found to be hearing impaired. A genetic component seems likely, given the high prevalence of hearing loss and the strength of family histories. However, we are limited in commenting on this issue since the cause of each individual's hearing loss was not established. Of the 60 children with a mild-moderate hearing loss, many would have been due to conductive hearing loss, which is almost entirely environmental in aetiology. Elahi et al[5] stated that 50% of hearing impairment, identified in children in rural Pakistan, was due to conductive hearing loss. Approximately 50% of childhood sensorineural deafness is 'genetic'[11]. Any 'genetic' hearing loss in our population would be non-syndroniic, there being no other congenital abnormalities mentioned.

Al-Gazali[12] showed that the majority of nonsyndromic, genetic hearing impairment is due to autosomal recessive inheritance. This form of inheritance will become exaggerated with consanguineous relationships. Although we found a high rate of consanguineous marriages, but no association was found between consanguineous parental relationship

and hearing impairment. Elahi et al[5] stated that in children with a severe hearing loss, 70% could be due to consanguineous marriages. Snashall[13] states that the usual prevalence of sensorineural hearing impairment is 1-1.5 per 1000. This figure may rise, to 12 per 1000 in populations where consanguineous marriages are common. Factors which may increase the prevalence further are quoted as: a positive family history, congenital abnormalities of the head and neck, prenatal and perinatal infections, prematurity, low birth weight, anoxia and hyperbilirubinaemia.

Many children identified will have a conductive hearing loss, It is likely that majority of these cases would have been due to otitis media, as evidenced by the link between a history of ear infections and hearing status, or otitis media with effusion (OME). The latter condition is probably more difficult to identify since it may run a fluctuating and asymptomatic course. It is important to identify as it may affect between 5 and 30% of children[14]. Nasal inflammation, resulting from allergic rhinitis or upper respiratory infections is a contributing factor to 0ME[15]. It would be of value to quantify the prevalence of' allergic rhinitis in Karachi, as children may be more at risk of OME in this environment than rural settings. Despite this message, the actual effects of otitis media on communication and educational development are yet to be fully established, with more prospective studies[16].

It is concluded, that routine screening of siblings of children with impaired hearing and an awareness campaign by hearing aid foundation, may be helpful in early detection and management of these cases.

This study has revealed an unacceptable level of hearing impairment in a population of children, whose parents are already aware of the importance of hearing loss. Some form of screening procedure must be implemented to curb this high rate of undiagnosed hearing loss. In light of our findings, it would be pertinent to routinely screen all siblings of children known to have a hearing impairment. Continuing work by the Ear Foundation Pakistan (EFP) can only improve the situation. However, in the absence of any screening program, we would further advocate an active campaign to heighten the public's awareness of hearing impairment. Hopefully this would improve parental suspicion and increase their demand for audiological testing.

References

1.Fortnum H, Davies A. Epidemiology of permanent hearing impairment in the Trcnt, Region. Br, J. Audiol., 1997,31:409-46.

2.Giles M, OBrien P. The prevalence of hearing inpairment amongst Maori school children. Clin. Otolaryngol., 1991,16:174-8.

3.Schlin P, Holmgren 0, Zakrisson J. Incidence, prevalence and etiology of hearing inpairment, in children in the country of Vasterbotten, Sweden. Scand. Audiol., 1990;19: 193-200.

4.Streppel M, Richling F, Walger M, et al. Epidemiology of hereditary hearing disorders in childhood: a retrospective study in Germany with special regard to ethnic factors Scand. Audiol.,2000;29:3-9.

5.Elahi MM. Elahi F, Elahi A, et al. Paediatric hearing loss in rural Pakistan. J. Otolaryngol., 1998;27:348-53.

6.Zakzouk SM, Bafaqech SA. Prevalence of severe to profound sensorineural hearing

loss in children having family members with hearing inpairment. Ann, Otol. Rhinol. Laryngol., 1996;105:882-6.
7.Davies A. Epidemiology of hearing inpairment. (Ch. 7: Diseases of the ear). 6th ed. Oxford, England, Oxford University Press, 1998.
8.Naeem Z, Newton V. Prevalence of sensonneural hearing loss in Asian children. Br. J, Audiol., 1996;30:332-9.
9.Finckh-KramerU, Spormann-Lagodzinski ME, Nubel K, et al. Is diagnosis of persistent pediatric hearing loss stilimade too late?, Hals-Nasen-OhrenHeilkunde, 1998;46:598-602.
10.Walkin PM, Baldwin M, Laoide S. Parental suspicion and identification of hearing inpairment Arch. Dis, Child., 1990;65:846-50.
11.Reardon W, Genetic deafness. J. Med. Genet., 1992;29:521-6.
12.Al-Gazali LI. A genetic aetiological survey of severe childhood deafness in United Arab Emiratcs. J. Trop. Pediatr., 1998;44:157-60.
13.Snashall S. Childhood Deafness. (Ch. tO: Diseases of the ear).6th ed. Oxford, England, Oxford University Press, 1 998
14.Daly KA, Hunter LL, Giebink OS. Chronic otitis media with effusion. Pediatr. Rev., 1999;20:85-93, quiz 94.
15.Fireman P. Otitis media and eustachian tube dysfunction: connection to allergic rhinitis. J. Allergy. Clin, ImmunoL, 1997;99:S787-97.
16.Gravel .TS, Wallace IF. Language, speech and educational outcomes of otitis media. J. Otolaryngeal., 1998;27(Suppl 2): 17-25.