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Original Article

Consanguinity and Hearing Impairment in a Deaf School

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Abstract

Objective: *To find out the frequency of consanguinity and positive family history of hearing impairment among deaf children in a deaf school in Bangladesh.*

Methods: *This was a retrospective study were included 150 patients. They were subjected to complete audiological examinations. These pupils were collected from Integrated Pre-school for Hearing Impaired children (IPSHIC) of SAHIC, Mohakhali, Dhaka, Bangladesh.*

Results: *The results showed that out of 150 Hearing impaired (HI) children 30.0% (45) were from the consanguineous parents and 70.0% (105) from non- consanguineous parents. Also found 22.0% (33) had positive family history of hearing impairment.*

Conclusion: *Prevention is only means to reduce the prevalence of Congenital hearing impairment.*

Key words: *Consanguinity, Hearing impaired, Deaf School*

Introduction

Deafness is the hidden disability of childhood, and leads to poor educational and employment prospects. Hearing impairment has debilitating effects on children as it can retard individual's language acquisition skills and impair the overall development.[1]

In UK approximately one child in 1000 is born with a bilateral permanent childhood hearing impairment (PCHI).This is defined as confirmed permanent bilateral hearing impairment exceeding 40dBHL(Hearing level) (average of pure tone thresholds at 0.5, 1, 2 and 4 KHz in the better hearing ear). About 60% of these children have a moderate (41-60dBHL) Hearing loss, while the remainder have a severe (61-80dBHL) or profound (>81dBHL) loss. WHO estimated in 2005 that there were 278 million people worldwide with bilateral moderate to profound hearing loss of whom 62 million had deafness that begin in child hood. 2/3rd of people with moderate to severe hearing impairment live in developing countries.[2]

In Bangladesh, deafness is a major public health problem. The country has a population of over 130 million and about 13 million people are suffering from variable degree of hearing loss which 3 million are suffering from severe to profound hearing loss leading to disability.[3]

In nearly half of children in the world with permanent hearing impairment the cause is

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genetic.[4] Of the genetic causes of hearing loss, syndromic form of deafness accounts 30% and non-syndromic hearing loss accounts for nearly 70% cases.[5] Major causes of deafness among children are genetic. Approximately 70-80% is autosomal recessive,15-20% autosomal dominant and 2-3% are x-linked or mitochondrial.[6,7] It is already established that 50 percent of the causes of deafness are preventable.[8] There is evidence that profound hearing impairment is more prevalent in countries where consanguineous marriages are common.[9]

Consanguinity refers to marriages between individuals who share at least one common ancestor and is conventionally applied to persons related as first or second cousins or to relatives within the extended family or same tribe.[10]

Consanguineous marriages are traditional in many Asian, African and South American communities. Consanguineous marriages are most prevalent in Arab countries, followed by India, Japan, Brazil and Israel. This is most common in lower educational and socioeconomic groups, among the traditionally religious, and in those marrying young. Particularly in the Muslim community, 20-50 percent of all marriages are consanguineous.[11]

The most recent consanguinity estimate indicated that some 10.4% of the world population are either married to a biological relative or are the progeny of a consanguineous union. There is no prescription that Muslims marry within the family, and first cousin marriage is freely permissible in the Jewish, Christian protestant, Buddhist and Zoroastrian / parsi religions. Also seen in the Dravidian Hindu populations of southern India preference is for a first cousin marriage between a man and his mother's brother's daughter, or more often marriage between an uncle and niece.[12]

In Pakistan, the prevalence of consanguinity is reported to be 60 percent.[13] Many studies have shown that the off spring of consanguineous marriages have a greater incidence of autosomal recessive diseases, including hearing impairment.[14] In addition ,the great majority of hereditary deafness is caused by single gene, autosomal recessive inheritance.[15]

Non-syndromic hearing impairment accounts for 80 percent of cases of hereditary deafness. Mutations in the same single gene have been shown to be responsible for both autosomal dominant and autosomal recessive forms of hearing impairment(e,g-connexin26).[16] A single locus, DFNB1, has been found to contribute to the majority of cases of autosomal recessive, non-syndromic hearing impairment.[17]

One study shows, the prevalence of deafness in children of Bangladeshi origin resident in East London up to 16 years of age was calculated to be at least 3.86 per 1000 which is 2.3 times higher the national average of 1.65/1000. In this study, the excess of childhood deafness was also attributed to genetic cause secondary to the practice of consanguineous marriage. One analysis suggests that consanguinity contributes to the raised prevalence of deafness in British Bangladeshi population. The distribution of causes of deafness has been genetic causes in 60%, acquired/ environment in 18%, and unknown in 22% of the 134 subjects in the study carried in East London Bangladeshi population. Amongst the genetic causes, most of the children (58%) had non-syndromic deafness. Even after genetic analysis, the cause of deafness remains undetermined in 22% patient.[18]

In developing countries, the WHO recommends that children's hearing should be screened at least at school entry, using

simple audiometer, and that the external ear be inspected for the presence of discharge. Earlier detection with universal neonatal screening is now practiced in developed countries.

In Bangladesh, the government runs seven deaf schools, where education is given free of cost. In the non-government sector there are about thirty schools for deaf.[3] Special teaching should be given by teachers of the deaf and available for deaf and partially hearing children.

Methods

This Retrospective study was carried out in Integrated pre-school for hearing impaired children (IPSHIC) of SAHIC, Mohakhali, Dhaka-1212.

Study period of February 2012 to July 2012 (6 months).

One Hundred and fifty (150) children was Purposely selected on single random method. The patient selected on the basis of Non probability purposive sampling method (Judgment sampling) who are admitted student in the IPSHIC. The parents or guardians of the patients were informed about the study and their consent was obtained.

Results

Table-I

Distribution of Type of marriage of parents of Hearing Impaired child by family history (n=150)

Marriage	Family History		
Consanguineous	Positive	Negative	Total
1st cousin	15	18	33
	45.5%	15.4%	22.0%
2nd cousin	4	8	12
	12.1%	6.8%	8.0%
Unrelated	14	91	105
	42.4%	77.8%	70.0%
Total	33	117	150
	100.0%	100.0%	100.0%

Of these 150 children, 33 (22.0%) had parents who were first cousin, 12 (8.0%) were second cousins. Out of 150 children 30.0% (45) were from the consanguineous parents and 70% (105) were from non-consanguineous parents.

Table-II

Distribution of Risk factors by family history (n=150)

Risk factors	Family History		Total
	Positive	Negative	
Consanguinity	12	33	45
	36.4%	28.2	30.0%
Family history	6	27	33
	18.2%	23.1%	22.0%
LBW & Prematurity	1	14	15
	3.0%	12.0%	10.0%
Birth asphyxia	0	9	9
	-	7.7%	6.0%
Neaonatal jaundice	0	6	6
	-	5.1%	4.0%
Meningitis	4	8	12
	12.1%	6.8%	8.0%
Maternal infection	7	5	12
during pregnancy	21.2%	4.3%	8.0%
Otoxic drugs	0	3	3
	-	2.6%	2.0%
No factors identified	3	12	15
	9.1%	10.3%	10.0%
Total	33	117	150
	100.0%	100.0%	100.0%

Others risk factors were LBW and prematurity (10.0%), meningitis (8.0%), Maternal viral infection during pregnancy (8.0%) and Birth asphyxia (6.0%).

Table-III

Distribution of Audio logical test for diagnosis of deafness (n=150)

Audiological test	Family History		Total
	Positive	Negative	
Pure tone audiometry (PTA)	15	63	78
	45.5%	53.8%	52.0%
Behavior observation Audiometry (BOA)	12	36	48
	36.4%	30.8%	32.0%
Free field audiometry (FFA)	6	18	24
	18.2%	15.4%	16.0%
Total	33	117	150
	100.0%	100.0%	100.0%

52.0% of deafness was detected by Pure tone audiometry.

Table-IV

Distribution of Degree of hearing loss (n=150)

Degree	Family History		Total
	Positive	Negative	
Profound (>90dB)	31	104	135
	93.9%	88.9%	90.0%
Severe (71-90dB)	2	13	15
	6.1%	11.1%	10.0%
Total	33	117	150
	100.0%	100.0%	100.0%

90.0% of the children presented with profound hearing loss and only 10.0% with severe deafness.

Table-V

Distribution of type of hearing loss (n=150)

Type of hearing loss	Family History		Total
	Positive	Negative	
Sensorineural	25	98	123
	75.8%	83.8%	82.0%
Mixed	8	19	27
	24.2%	16.2%	18.0%
Total	33	117	150
	100.0%	100.0%	100.0%

Majority of patients hearing loss (82.0%) presented were sensorineural type.

Table-VI

Distribution of Type of using Hearing device (n=150)

Hearing device	Family History		Total
	Positive	Negative	
Body worn	21	66	87
	63.6%	56.4%	58.0%
Behind the ear	11	37	48
	33.3%	31.6%	32.0%
Cochlear implant	1	14	15
	3.0%	12.0%	10.0%
Total	33	117	150
	100.0%	100.0%	100.0%

Majority of children uses Body worn type of hearing aid (58.0%) and a number of deaf children used Cochlear implant.

Discussion

In this study 150 deaf children aged below 12 years have been studied. Emphasis was given on history, clinical examination and audiological investigation. Although this study had been carried out within a limited period of time and with a limited number of patients yet this may in some extent reflex the overall situation of the society as the patient had been collected from a referral from different areas of the country. In this series, age distribution of the children presented to the deaf school showed that maximum number patients (60.0%) were in the age group between 5-8 years.

This study reflected parents were consanguineous in 30.0% (1st cousin 22.0% and 2nd cousin 8.0%), which is supported by the study Bajaj Y et al.[18] who also found consanguinal marriage in 33% in British-

Bangladeshi community. Various other studies conducted amongst children born to consanguineous parents reveal a much higher incidence of hearing loss in this group.[9, 10,11]

This study also reveals a family history of deafness in 22. 0%, this is supported by the study J.O. Hara et al[19] also found positive family history of deafness 26%.

Others risk factors of deafness identification was mainly based on history taken from the patient's parents , LBW and prematurity (10.0%), Meningitis(8.0%), Birth asphyxia (6.0%) which was supported to the study by Minja BM at el. [20] except Maternal viral infection during pregnancy(8.0%).

BOA) was done in 32.0% and free field audiometry (FFA) was done in 16.0% of deaf children. Here Pure tone audiometry (PTA) was done 52.0%, behavior observation audiometry .90.0% of deaf children presented with bilateral profound hearing loss and 20.0% presented with bilateral severe hearing loss. 82% were bilateral sensorineural and 18% were bilateral mixed type hearing loss . The above results are consistent with findings of that series.[21]

This study shows that the 150 deaf child were managed with Hearing aid and Cochlear implant, among them 58.0% uses Body worn (BW) hearing aid, 32.0% uses Behind the ear (BTE) & 10.0% uses Cochlear implant (CI). Most of the child 54.0% uses hearing aid in both ears, 26.0% uses in Rt. Ear and 20.0% uses in Lt. ear. WHO report of an inter-country consultation showed that Body worn (BW) type was dominant in India, where as BTE type in Indonesia.[22]

In this study population, Consanguinity 30% was observed. This high percentage of consanguinity indicating that the consanguinity is a risk factor for hereditary hearing loss, but this is not overall focus in Bangladesh.

This subject also needs further investigation to rule out genetic causes such as GJB2 gene (connexin 26) mutation etc. [23]. However such testing was beyond the purview of the present study. This protocol should include both, audiological diagnostic test as well as suitable immunological and genetic studies to establish the causes of the condition.

Conclusion

Condition such as Consanguinity require more research to establish or discredit them as potential causes of childhood hearing loss. Prevention is only means to reduce the prevalence of congenital hearing impairment. This can be achieved by genetic counseling of individually and families at risk. Further large number of hearing impaired babies gave history of conditions that are preventable, such as Low birth weight, Birth asphyxia, meningitis etc. This suggest that a large number of babies could be prevented from developing hearing loss.

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