Consanguinity as Cause of Hearing Loss :A study of 200 Cases

Imad, Khairulaman, Sadia Farhad

ABSTRACT

Background: Consanguinity is believed as marriage between cousins or more closely related family members. It is considered an important causative agent in the prevalence of hereditary disorders. Consanguinity has been reported being more common among parents of children having hereditary hearing loss.

Objective: To study the frequency of consanguinity in hearing impaired children in District Mardan at Otorhinolaryngology department of Mardan Medical Complex teaching hospital Mardan.

Material and Methods: It was a prospective study on 200 children with age range of 2-5 years, visiting otorhinolaryngology department for hearing impairment. Only patients with hereditary hearing loss were included in the study. Patients with deafness because of other reasons like birth asphyxia, prematurity, head injury etc. were excluded from the study. A thorough history, complete audio logical examination and investigations were done.

Results: Out of 200 cases, consanguinity was found in 67.5%, while 32.5% were non consanguineous . In 98 cases parents were 1st cousins while in 37 cases parents were 2nd cousins. 78% of patients had profound senserineural hearing loss while 28% had severe senserineural hearing loss.

Conclusion: Congenital hearing impairment can be reduced by discouraging and counseling against consanguinity. Consanguinity is an important community health problem and is the major cause of hereditary hearing loss in children.

***Key words**: Consanguinity , Etiology , Hearing loss*

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INTRODUCTION

Hearing loss is a major cause of referral to outpatient department of ENT it is found in all ages and both sexes. Hearing impairment has strong adverse effects on individual's learning, and overall mental and physical development[1]. Deafness is one of the rapidly rising disabilities among human beings and responsible for one third of the entire disease burden in the world[1]. Causes of hearing loss may be congenital, perinatal or acquired. Prevalence of congenital profound hearing loss worldwide is 11 per 10,000 children and is said to be due to genetic causes in at least 50% of the cases. One in 1000 newborns in UK are reported of having bilateral permanent childhood hearing impairment[2]. It may be monogenic or diagenic. Congenital hearing loss is not always occurred by birth, the child may inherit the tendency to develop it later in life[3].Senserineural hearing loss whether hereditary or environmental, occurs 1-3 of every 1000 children of developed part of the world and it may be more than this in under developed countries[4]. A hearing impaired child has limited exposure to environmental sounds and speech which prevents normal auditory developments[4].

1. Bacha khan Medical College Mardan

Correspondence: Dr. Imad
Associate professor ,E N T ,
Bacha khan Medical College Mardan.
Cell no. 03339155434
E.mail .imad.hameed@yahoo.com

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Consanguinity has been attributed as a major factor towards congenital hearing loss[5-8] . It is among one of the preventable causes[9]. Consanguinity is defined as marriage of parents with a recent common ancestor, who may give the child the same recessive gene received from the same ancestor. Consanguinity as of genetic significance was first studied by Garrad in 1902 [10] .

Consanguineous marriages are very common in our society especially in the tribal belt of KPK and Balochistan. Though it is mostly because of local customs but socioeconomic status also plays a major role like inheritance of family properties and poverty etc.[11]

It is not only the Muslims but Jewish, Christian protestants, Buddhist and Parsi religions also prefer marriages within the family. The recent consanguinity estimate all over the world is 10.4%[12]. In Pakistan the prevalence of consanguinity is reported to be 60 percent[13].

The purpose of the study was to evaluate the role of consanguinity in hearing loss among children.

MATERIAL AND METHODS

A prospective study was carried out on children with age range of 2 to 5 years, with hearing impairment at ENT department of otorhinolaryngology Mardan Medical Complex teaching hospital Mardan.

This was a Prospective observational study on 200 children with hereditory hearing loss . The duration of the study was 1st July 2014 to 13th June 2017.

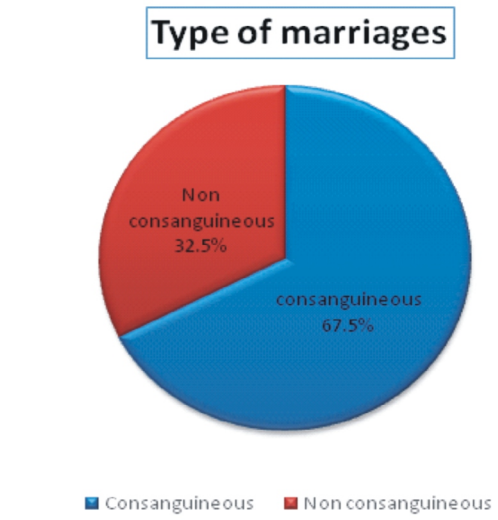
A questionnaire was made to collect all the necessary information. Information was obtained from parents and individual records. A complete medical, family ,prenatal ,natal and post natal history was taken. Complete ENT examination and audiological investigations were carried out to find the type and degree of hearing loss. In case of no high risk environmental factors and family history of deafness in close relatives, the deafness was considered congenital.

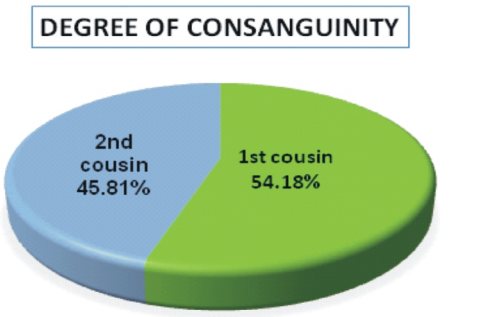
Only cases of congenital hearing loss were included in the study. Hearing loss due to non genetic environmental factors like rubella, prematurity, birth asphyxia, meningitis, kernicterus ,and head injury were excluded from the study.

Consanguineous marriages in our study were the ones in which parents had atleast one ancestor in common and the ancestor is no more distant than a great grandparent.

RESULTS

In this study of 200 children with hereditary hearing loss, consanguinity was found in 67.5% (135 out of 200) of cases. 32.5% (65 out of 200) cases were non consanguineous (table-1).In 54.81% (74 out of 200) cases parents were 1st cousin and in 45.18% (61 out of 200) parents were 2nd Cousin(table-1). Profound hearing loss was seen in 78% and severe hearing loss was noted in 28% of children affecting quality of life adversely(table-1).





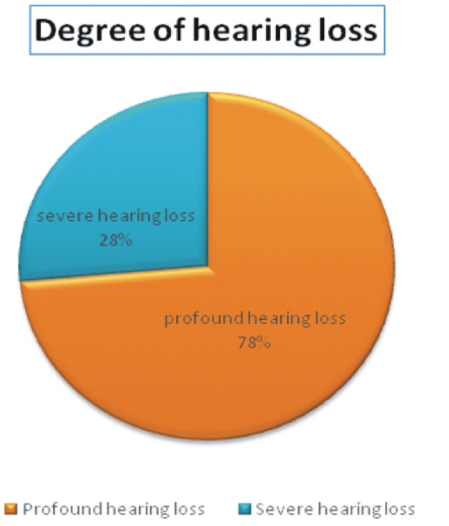


Table No. 01

Types of Marriages		Degree of Consanguinity		Degree of Hearing loss	
Consanguineous	Non consanguineous	1st cousin	2nd cousin	Profound hearing loss	Severe hearing loss
135 (67.5%)	65 (32.5%)	74 (54.81%)	61 (45.18%)	105 (78%)	30 (28%)

DISCUSSION

Consanguinity has been established as a high risk factor for hereditary hearing loss in various communities of the world. Being more common in Asian and African countries, it has also been found that consanguinity in hearing impaired children is more than general population.

In our study consanguinity was found in 67.5%of the hearing impaired children ,which is more than the 60% of consanguinity in general population. This is in accordance with Ajallouyan study who reported consanguinity in 65% of parents of deaf children which is much more than 38% of consanguinity in general pollution of Iran[4]. Saadat also found 38.6% of consanguinity in Iran with 27.9% as first cousin marriage[5]. Reddy reported consanguinity in general population of Andrapardesh India to be 22.36% but this was 41.73% in deaf children of his study[1].Consanguineous marriages cause an increase in the homozygosis of mutated recessive genes resulting in birth of hearing impaired children.

Debate came across 30% cases of consanguinity in his study[12].This is comparable with the study by Bajaj et al who reported cousin marriages in 33% of Britian-Bangladeshi community[15]. Though khabori et al reported that 70% of the deaf children were from parents of consanguineous marriages[16].Amini et al showed 61.4% hearing impairment in cousin marriages[17].

In our study consanguinity as 1st cousin marriage was seen in 54.81% and second cousin marriage in 45.18% of cases. This is supported by Ajallouyan who found 1st cousin marriage in 42.5%, though Zakzouk reported 1st and 2nd cousin marriage in 22% and 23% of deaf children respectively.

CONCLUSION

Consanguinity is an important community health problem and is the major cause of hereditary hearing loss in children. 1st cousin marriage as consanguinity is one of the major factors.

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