

Sensorineural Hearing Loss seen in Siblings of Consanguineous Marriages at Multan

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ABSTRACT

Aim: To identify the role of consanguinity in the appearance of deafness in children of consanguineously married couples.

Materials & methods: This retrospective, record based, survey type study was carried out during April-July, 2007. We completed a questionnaire for each student after interviewing the student and the parent or guardian. We also consulted the record at the school. The data was analyzed to see the frequency of consanguinity among parents of hearing impaired children.

Results: It was seen that 65.60% parents of children with congenital sensorineural deafness, were first cousins; 15.20% parents were second cousins and 19.20% of parents were not related to each other. A high percentage (63.20%) of the participants had some other member of the family having the same type of hearing abnormality.

Conclusion: Consanguineous marriages seem to be associated with higher incidence of sensorineural hearing loss among their siblings.

Key words: Consanguinity, sensorineural hearing loss, congenital deafness

INTRODUCTION

Consanguinity literally means a relationship based on descent from a common ancestor or a relationship by blood. By consanguineous marriage we simply refer to marriages between such males and females who are related to each other by blood. The study of close kin marriages is important to look into its consequences in terms of fertility, child morbidity and child mortality¹.

In the 21st century also, consanguineous marriages remain popular in many parts of Asia and Africa and it has been estimated that currently more than 10% of the global population are either married to a partner related as second cousin or closer ($F \geq 0.0156$) or are the progeny of such a union².

In Pakistan, marriages between spouses related as second cousins or closer accounted for 50.30% of the total marriages³. In another study⁴, Hussain and Bittles (1998) showed that in Pakistan, 60-80% of marriages are among first cousins and this trend remained unchanged over the past 3-4 decades.

Hearing impairment even of a mild degree can result in long lasting communicative, social and academic deficits. It is well documented that presence of hearing loss will interfere with the acquisition, development and the use of language⁵. Congenital sensorineural hearing loss is an autosomal recessive condition which is allegedly

seen in increasing proportions among those who commonly practice consanguinity⁶.

Consanguineous marriage is a tradition which is commonly practiced among Asian, African and Latin American communities whether they are living in their own countries or settled in Europe or the USA. These communities, in addition to their custom of interrelated marriages, have large families and are a rapidly growing population. The siblings of consanguineous marriages have a significantly higher incidence of autosomal recessive diseases including hearing impairment⁷.

Hereditary hearing impairment is genetically heterogenous group, exhibiting patterns of inheritance that include autosomal recessive, autosomal dominant, X-linked and mitochondrial. Nonsyndromic deafness represents about 70% of all cases. Nonsyndromic autosomal recessive deafness accounts for about 80% of nonsyndromic hereditary hearing loss. To date, 52 loci responsible for this type of deafness are mapped and 24 genes identified⁸.

In the present epidemiological study, we tried to identify the role of consanguinity in the appearance of deafness in children of consanguineously married couples.

SUBJECTS AND METHODS

This retrospective, record based, survey type study was carried out in the Department of Physiology, Nishtar Medical College, Multan in collaboration with the Special Education Institute for Deaf and Dumb,

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After taking permission from the Head of the Institution, and proper consent from the parents of the special children, we completed a questionnaire for each student. The questionnaire contained information regarding age, sex, consanguinity of parents, deafness among other family members and definitive diagnosis of the type of deafness on the basis of the ENT specialist prescription. We inspected the records of the students admitted to the Institute, interviewed the students through an interpreter and where possible we also interviewed the parents or guardians. All the students, whose medical and other records were complete, were included in the study.

Data were analyzed to see the frequency of type of deafness, effect of consanguinity of parents and family history of deafness. Effect of consanguinity was assessed by noting the frequency of deafness seen among siblings of consanguineously married couples and near relatives.

RESULTS

Important findings of the study are shown in the given table. Among 250 consenting and eligible participants, 150 were males and 100 were females. The age range of the participants was 08-18 years. Among males 89.30% and among females 98.00% were congenitally deaf. Acquired deafness was seen among 10.70% male and 02% female participants. About parental consanguinity, it was seen that 65.60% parents of the participants were first cousins, 15.20% parents were second cousins and 19.20% of parents were not related to each other. A high percentage (63.20%) of the participants had some other member of the family having the same type of hearing abnormality.

Table: Important Findings of the Study (n=250)

Characteristics	Frequency
Baseline Characteristics	
Males(n=150)	60%
Females(n=100)	40%
Age Range(years)	08-18
Congenital deafness	
Males(n=134)	89.30%
Females(n=98)	98%
Acquired Deafness	
Males(n=16)	10.67%
Females(n=02)	02%
Parental Consanguinity	
First cousin Parents(n=164)	65.60%
Second cousin Parents(n=38)	15.20%
No Relation Parents(n=48)	19.20%
7Deafness in Other Relatives (n=158)	63.20%

DISCUSSION

This study was aimed to find out whether consanguinity in marriages is a risk factor for the hearing impairment in children of Multan District. As consanguinity is common among Muslims all over the world and in Pakistan different sects and linguistic groups of Muslims tend to marry within their religious bonds, it was relevant to study this relationship in the local community⁶.

Consanguinity is practiced throughout the world e.g., North and Sub-Saharan Africa, West, Central and South Asia has 10-50% ratio of consanguinity among general population. In Japan, South America and Siberian Peninsula this is 01-10% and in Western Europe Australia and Russia this is only 01% in general population. In Pakistan it is reported to be around 60%². Consanguineous unions are now either illegal or a criminal offense in 31 of 50 states of USA^{9,10,11}.

In our study, consanguinity among parents of hearing impaired was 65.60% (first cousin parents), and 15.20% (second cousin parents). Our findings are similar with the findings of Abolfotouh and Al-Ghamdi (2000) who reported same percentage of consanguinity among parents of their study population⁸. As reported earlier⁷, the incidence of hereditary hearing impairment is very high in developing countries, the frequency seen in present study seems to be one of the highest. Preventive measures are to be adopted early and so, it is recommended that in our population consanguinity in marriages should be discouraged.

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