Hearing impairments in consanguineous marriage

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Abstract:

Consanguineous marriage is strongly favored in many large human populations. In the most parts of south Asia, consanguineous marriage account for 20% to over 50% of the general population .

The effect of consanguinity on hereditary deafness has been well studied and documented. Many authors have suggested that approximately one half of sensory neural hearing loss in children can be attributed to hereditary causes.

This research was carried out in Rehabilitation Deputy of welfare organization of Iran in seven provinces. 1352 infants and preschool aged children participate in this research. The prevalence of SNHL due to consanguineous marriage in first cousin and second cousin were studied.

Consanguinity was found among 45.7 percent in first cousin and 17.2 percent in second cousin. Hereditary Factors were thought to be the cause of 863 (62.9 percent) of bilateral SNHL children in this research.

The incidence of hereditary hearing impairment is very high in developing countries compared to developed countries . Prevention is essential to reduce the incidence of genetic hearing loss. Consanguinity should be discouraged and genetic counseling is to be applied at least for those at risk of developing genetic diseases including hearing impairments.

Key words :

Consanguineous marriage, hearing impairmens , sensory-neural hearing loss

Introduction :

Consanguineous marriage occurs in varing degrees throughout the world. It is particularly prevalent in some part of Middle East, Asia, African and Latin American communities (1-2).

In the most parts of south Asia, consanguineous marriage accounts for 20% to over 50% of the general population .

The siblings of consanguineous marriages have a significantly higher incidence of hereditary disease including hearing impairments (4). The risks are greater, too, in families which have a genetic disorder in this case, marrying a relative can lead to a much higher risk of having an affected child.

The great majority of hereditary deafness is caused by single autosomal recessive inheritance(5) .The effect of consanguinity on hereditary deafness has been well studied and documented.

Many authors by using statistics principles and causes of deafness believe that consanguineous marriage will increase the chance of deafness by autosomal recessive genes. The purpose of this research was to study the prevalence of SNHL due to consanguineous marriage (the hereditary type) and to review the literature and the possi-

bilities of preventing or minimizing this custom.

Material and Methods :

In this research , 1352 infants and pre-school age children, between ages 1 month to 6 years, were assessed during February 2001 to October 2002 . This study was performed in rehabilitation centers of Iran Welfare Organization (I.W.O) in seven provinces (Tehran , Esfahan, East Azarbayejan , Khorasan , Khoozestan , Mazandaran and Fars) .
All of children had bilateral symmetrical sensory - neural hearing loss.
A questionnaire was prepared and completed with the help of the parents. It consisted of information regarding age, sex, consanguinity of parents, family history of deafness, hearing and speech deficits and exposure to various known risk factors for hearing impairment.
A child was considered to have hereditary hearing impairment if there was a positive family history. A sibling was considered as having a positive family history if a parental sibling of direct parent ancestors was deaf without environmental factors, regardless of the parental hearing status. Children with positive environmental factors (prematurely, hyperbilirubinemia, meningitis, rubella, etc) were considered to be deaf as a result of these factors.

Table I

Age and Sex Distribution of 1352 Children

Age Category	*Number*	*percent*
0-12 month	25	1.8
13-24 month	98	7.2
25-36 month	243	18
37-48 month	269	19.9
49-60 month	274	20.3
61-22 month	443	32.7
Total	1352	100
Sex		
Female		49.1
Male		50.9

Results :

In this survey 1352 Cases , 50.9 percent male and 49.1 percent female, were evaluated , Table 1 shows the age and sex distribution of the children .

Table 2 shows the relationship of the parents in each of provinces. In general the relationship of the parents were 631 (45.7 percent) in first cousin , 232 (17.2 percent) in second cousin and 501 (37.1 percent) in not related parents .

The data in table 2 shows that consanguineous marriage between first cousin is much higher in Khoozestan rather than other provinces in Iran (south of Iran) .

Hereditary factors were thought to be the cause of 863 (62.9 percent) of bilateral sensory neural hearing loss.

The prevalence of hearing impairment was founded to be significantly higher in the children whose parents were first cousins compared to the children whose parents were second cousins .

Table II

Relation of the parents in each of province

Province	Number	First cousin related	Second cousin related	Not related
Tehran	547	(42.9%)	(17.6%)	(39.5%)
Esfahan	150	(46%)	(16.7%)	(37.3%)
East Azarbyejan	88	(54.6%)	(5.7%)	(39.6%)
Khorasan	132	(41.6%)	(17.5%)	(40.9%)
Khoozestan	166	(60.8%)	(18.1%)	(21.1%)
Mazandaran	161	(37.1%)	(16.6%)	(46.3%)
Fars	111	(47.7%)	(25.3%)	(27%)
Total	1352	(45.7%)	(17.2%)	(37.1%)

Table 3 shows the degree of bilateral SNHL in children. 50.6 percent of children had profound hearing loss.

Table III

Degree of bilateral SNHL

Degree	*Number*	*Percent*
Slight	8	./6
Mild	22	1.6
Moderate	154	11.4
Moderate- severe	149	11
Severe	335	24.8
Profound	684	50.6
Total	1352	100.0

The prevalence of hearing impairment in the children and consanguinity of parents illustrate in figure 1 .

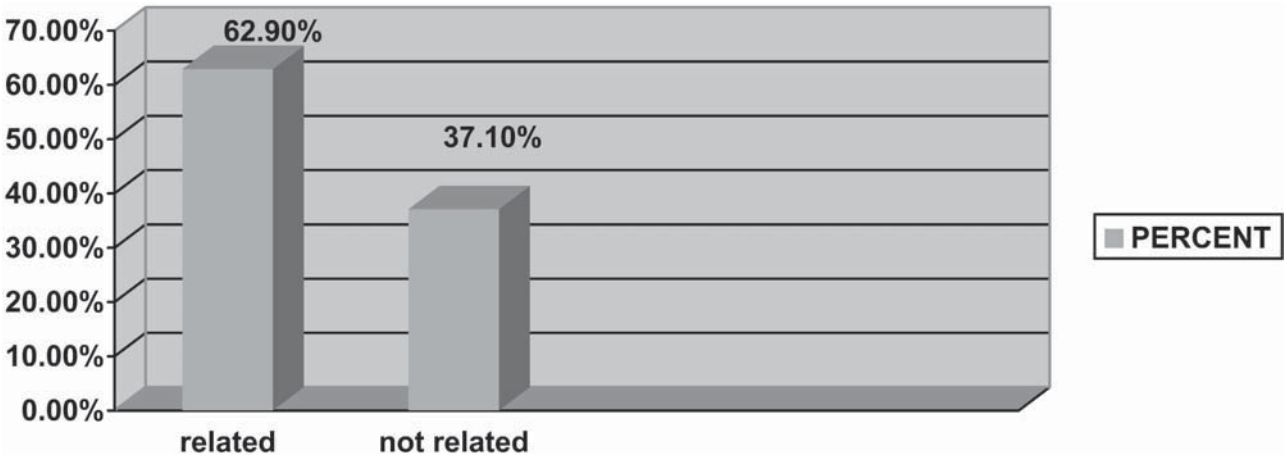
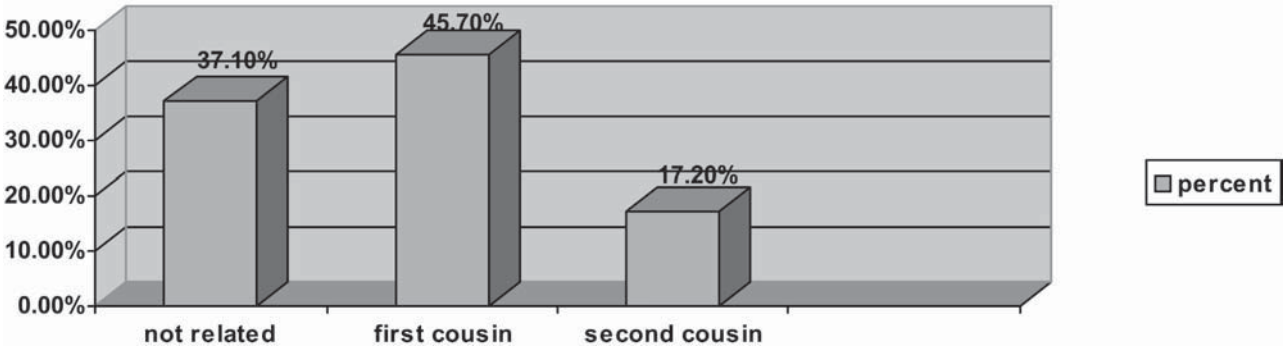


Figure 2 shows frequency distribution percentile of total subjects according to consanguinity of parents.



Discussion :

Consanguineous marriage is strongly favored in many large human populations . It can vary quite widely between and within countries, religions and cultural factors play a major part in determining social attitudes and legal frameworks at local and national levels. Some authors suggest such marriage should be avoided as they make double the risk of infants with birth defects such as mental retardation, deafness and blindness when compared with an "unrelated" marriage.

Many authors have suggested that approximately one half of sensory neural hearing loss in children can be attributed to hereditary causes (6). In this study hereditary deafness accounted for 62.9 percent of 1352 children with bilateral sensory neural hearing loss. The parents of 17.2 percent children were second cousins and 45.7 percent were first cousins .

Siraj Zakzouk (2002) reported 66.1 percent hereditary deafness in 168 children with sensory neural hearing loss in his first study and 36.6 percent hereditary deafness in 142 in his second study. He had explained this big difference in the prevalence of hereditary deafness between the two samples was due to increased awareness of the families and the improved health services (7). Taylor et al. (1975) reported that most , if not all, cases of deafness previously classified as of "unknown cause" are cases of autosomal recessive inheritance (8).

In the majority of cases of hereditary deafness (75 to 88 percent) inheritance is by a recessive single gene .

Consanguineous marriage also increases the risk of transmission of polygenic (multifactorial) inheritance . This uncommon type of inheritance is not fully understood, but it is postulated that multiple genes contribute to the disease and each individual has a threshold that about which the abnormality will be manifest (9). An important point is that for multifactorial inheritance, the risk to subsequent siblings is higher when the parents are consanguineous than when they are unrelated, in contrast to

autosomal recessive inheritance, where the risk is the same whether or not the parents are consanguineous .The effect of consanguinity on the development of childhood hearing impairment depend on the closeness of the relationship of parents. A marriage between first cousins poses a great risk, whereas a distant consanguinity has comparatively low risk of producing defective offspring, which is also supported by our findings.

The hazards of blood marriage and the mechanisms that contribute to it must be explain for the families . Genetic counseling does provide those seeking information or advice a true picture of the situation with its associated risks , so the family can take the appropriate decisions about marriage.

A preventive program is necessary to limit the number of children affected through public health education regarding the possible outcome risks of consanguineous marriage. Screening to identify carries of genetic disorders is an essential aspect of prevention. School screening , premarital, prenatal as well as neonatal screening should be part of the program .

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