

Consanguinity Among Parents of Iranian Deaf Children

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Abstract

Background: It seems that there is a relationship between consanguinity and profound hearing loss but there is little data about the association of consanguinity and hearing loss in Iran.

Objectives: The aim of this study is to demonstrate the causes of profound bilateral sensorineural hearing loss among Iranian samples who are candidates for cochlear implantation.

Methods: This study was retrospective, analytical, and designed to collect information about profound hearing impaired cases referred to the Baqiyatallah Cochlear implantation center using enumeration. A total of 310 children with profound hearing impairments participated in this study. They were aged from 6 months to 4 years old. The study was done between January 2007 and April 2009. Chi-square tests were used to show whether there was any statistical difference between the incidence of marital consanguinity of their parents and the normal population.

Results: Sixty-five percent of those 310 children had parents who had married with their relatives. Of the 203 (65%) parents that had consanguineous marriages, 132 were first cousins, which includes the children of two brothers (37 [11.8%] patrilineal parallel cousins), the children of two sisters (38 [12.2%] multi-lateral parallel cousins), or the children of a brother and a sister (57 [18.3%] cross cousins). Fifty-four (17.4%) of the parents were second cousins and 17 (5.2%) were beyond second cousins. Also, hearing loss etiology was obvious in 237 (76.3%) of the patients with profound hearing loss but was unknown in 73 (23.7%). Hereditary was identified as the most common cause in 33% of the cases.

Conclusions: Our data demonstrated a 65% occurrence of consanguineous marriage among the parents of deaf children, which is statistically different from the percentage of consanguineous marriage among Iranian population (38%). This indicates an obvious relationship between severe hearing loss and consanguineous marriage.

Keywords: Consanguinity, Hearing Loss, Children, Cochlear Implantation

1. Background

Sensorineural hearing loss (SHL), whether hereditary or environmental, occurs in 1 - 3 of every 1,000 children born in developed countries (1, 2), but this rate is probably higher in developing countries (3). Deafness limits learning and social communication, both of which are necessary for the normal development and maturation of a child. A deaf child receives little or no access to environmental sounds and speech, which disrupts normal auditory development (4-6).

Some studies estimate that about half of the deaf individuals have hereditary deafness (7-9). Consanguineous marriages, defined as marriage between individuals who are second cousins or more closely related cousins (10), are an important etiologic factor in the prevalence of genetic disorders. It is estimated that, globally, over 20% of

the human population and over 8.5% of all of the children have consanguineous parents (10). This is most prevalent in Middle Eastern countries, where the proportion of consanguineous marriages has been reported ranging from 20% to 70% (11). In Iran, it was reported in 38% of all marriages (12, 13). A significant relationship is reported between SHL and consanguinity by some authors (14-17), and it is reported that the etiologies of hearing loss can be related to consanguinity. The studies show that consanguineous marriage is higher in people who have children with hereditary hearing loss. Therefore, consanguineous marriages can result in hereditary hearing loss (14-17).

However, some studies demonstrate that the relationship between SHL and consanguinity is due to other etiologic factors such as febrile convulsion, meningitis, hyperbilirubinemia, and rubella. It seems that consanguineous marriage is decreasing; the decline is likely to be greatest

in the large cities (1-3).

2. Objectives

Although there have been studies in other countries on consanguineous marriages, there is little data about the relationship between consanguinity and profound hearing loss in Iran. Therefore, the aim of this study is to document the causes of profound bilateral SHL among Iranian patients who are candidates for cochlear implantation.

3. Methods

3.1. Subjects

This study is retrospective, analytical, and designed to collect information about Iranian profound hearing-impaired patients that were referred from all areas to the Baqiyatallah Cochlear implantation center. The sample size of the study was by enumeration according to the inclusion and exclusion criteria and the study of available files. A total of 310 children with profound hearing impairments from the Baqiyatallah Cochlear implantation center participated in this study. There were aged from 6 months to 4 years old.

3.2. Data Collection

A uniform questionnaire was used for all of the SHL cases. The details were filled in by the ENT surgeon. The study data was collected between January 2007 and April 2009. The form included the demographics and the other details relevant to the hearing impairment. This information was obtained from the parents, instructors, and individual records. The data included a complete medical and family history and prenatal, natal, and, postnatal high-risk factors. Also, non-genetic environmental factors like rubella (prenatal), prematurity, birth asphyxia, kernicterus (perinatal), meningitis, head injury, consumption of ototoxic drugs, or other infectious conditions (postnatal) causing hearing impairment were collected. A complete ENT clinical examination was performed that included general and systemic examinations of any congenital abnormalities.

The careful physical examination included systemic, otologic, and opthalmologic evaluations and blood count, urinalysis, and thyroid function tests (T3, T4, and TSH). Audiologic investigations were conducted to evaluate the degree and type of hearing loss. Brainstem-evoked response audiometry (ABR) was used to confirm the hearing impairment. If there were no high risk factors and the child had close relatives who are deaf, the cause of deafness was decided to be hereditary.

In this study, a consanguineous marriage represents one in which the two parents have at least one ancestor in common, with the ancestor who is no more distant than a great-grandparent. For descendants who were of the same generation, a consanguineous marriage was between third cousins or closer relatives. Consanguineous marriages were classified by the degree of relatedness between the parents: first cousins, second cousins, and beyond-second cousins. For a given degree of relatedness, a second level of classification was noted. First cousins may either be the children of two brothers (patrilateral parallel cousins), of two sisters (multi-lateral parallel cousins), or of a brother and a sister (cross cousins).

3.3. Statistical Analysis

We use SPSS version 15 and a Chi-square test to show whether there was a statistical difference between the incidence of marital consanguinity in deaf children and the normal population.

4. Results

Of the total 310 patients, 164 (53%) were male and 146 (47%) were female. The mean age of the children was 4.1 ± 1.87 years old. Of all the parents, 203 (65%) had consanguineous marriages and 107(35%) did not.

Of the 310 children, 103 (33%) had one or more close relatives who were deaf in their families; thus their cases were considered to be hereditary deafness. A total of 203 (65%) of the children had parents who had married their relatives. Of this, 132 of the parents were first cousins, that included the children of two brothers (37 [11.5%]), the children of two sisters (38 [12.2%]), and children of a brother and a sister (57 [18.3%]). Fifty-four (17.4%) of the parents were second cousins and 17 (5.2%) were beyond second cousins (as shown in Table 1).

As shown in Table 2, the etiology was obvious in 237 (76.3%) of all of the patients with profound hearing loss but was unknown in 73 (23.7%). Hereditary was identified as the most common cause in 33% of the patients. After the hereditary factor, febrile convulsions were the next-highest with 15.8%. Also, the Chi-square test shows that 65% of the patients' parents had consanguineous marriages; this rate is higher than the average of consanguineous marriages (38%) in Iran ($P < 0.0001$). Of these marriages, the parents of 49 (37.6%) children were married to their first cousins and 15 (11.5%) were married with other close relatives.

Also, a systemic examination shows behavioral disorders in 153 (49%) of the patients and a developmental delay in 42 (32.3%) under the age of 1. Between the ages of 12 and 30 months, 52 (40%) of the children with hearing loss were

Table 1. Percentage of Consanguineous Marriages in Parents of Children with Hearing Loss

Consanguineous Marriages	Numbers of Patients (%)
Patrilateral parallel cousins	37 (11.5)
Multilateral parallel cousins	38 (12.2)
Cross cousins	57 (18.3)
Second cousins	54 (17.4)
Beyond second cousins	17 (5.2)
Total	203 (65)

Table 2. Causes of Hearing Loss in Children

Causes	Numbers of Patients (%)
Unknown	73 (23.7)
Hereditary	103 (33)
Febrile convulsion	49 (15.8)
Meningitis	18 (5.8)
Hyperbilirubinemia	9 (3)
Rubella	9 (3)
Measles	8 (2.6)
Prematurity	8 (2.6)
Oto-toxicity	8 (2.5)
Birth trauma	8 (2.6)
Chronic otitis media	7 (2.2)
Trauma	4 (1.3)
Encephalitis	2 (0.6)
History of CMV in mother	4 (1.3)
Total	310 (100)

diagnosed, while 36 (27.6) of all cases could not be identified by the age of 30 months.

5. Discussion

Our data shows the prevalence of consanguineous marriage among the parents of deaf children (65%). The overall rate of consanguineous marriages in our patients is much higher than the rate in the normal population. These findings show the importance of consanguinity as a predisposing factor for child deafness.

Consanguinity is prevalent in some areas of Asia and Africa because of socioeconomic, cultural, and religious factors (18, 19). Historical studies report a strong preference of Iranians for consanguineous marriages (12, 13). Due to the Islamic rules mentioned in the Holy Quran, men are prohibited to marry their close relatives (e.g., a man's

mother, sisters, daughters, aunts, nieces, and some of his wife's relatives), but are allowed to marry with any of their parallel or cross cousins (18). The mean proportion of consanguineous marriages in Iran is 38 % in the normal population, ranging from 15.9% in the Northern provinces to 47.0% in the eastern provinces (12, 13, 20).

First-cousin marriages are the most common form of consanguineous marriage in the parents of our patients. The rates of first-cousin, second-cousin and beyond-second-cousin consanguineous marriages were 42.5%, 17.3%, and 5.2%, respectively. In first-cousin marriages, the cross-cousin type was higher than patrilateral and multilateral parallel cousins. In fact, a general bilateral preference for marriage with close kin has been reported in many regions of Iran (12, 20, 21). It can increase the frequency of all autosomal recessive disorders. Moreover, the gene of autosomal recessive disorders in the community

is expressed more in first-cousin marriages (10).

Previous studies on etiologic factors of deafness in different countries suggest that approximately one half of SHL in children is attributed to hereditary causes (21, 22). Our findings match this. In a study from Saudi Arabia, reported by Zakzouk et al (2002), hereditary deafness occurred in 66.1% of 168 children with SHL (17, 22, 23). This relatively high incidence of hereditary deafness may be explained by the actual increase of hereditary deafness cases due to consanguinity and/or of the improvement of detecting genetic deafness. The Bener et al. (2005) (24) study from Qatar showed that parental consanguinity was more common among HL cases (60.5%).

The effect of consanguinity on the genetic origin of deafness has been demonstrated in other parts of the world (24). In a retrospective survey, Feinmesser et al. (1966) (25) showed consanguinity to be more frequent in 224 deaf children of Jewish origin than the normal population. Bergstrom et al. (1971) (26) demonstrated that the marriage of two deaf people gives only a slightly increased risk of deafness in the children because there is a small chance that two such people would be affected by the same genetic deafness. On the other hand, if the parents are consanguineous, they are more likely to be homozygous for the same trait and, therefore, all their children will be affected. Ansar et al. (2003) (16) reported the localization of an autosomal recessive, non-syndromic hearing impairment locus (DFNB38) to 6 q 26 q 27 in consanguineous parents from Pakistan. The study illustrates the high prevalence of hereditary deafness and consanguineous marriage among their population. In a study from Iran, Amini et al. (2010) (27) showed that the prevalence of hearing impairment in consanguineous marriage was 61.4%.

Prevention is the only way to reduce the incidence of genetic hearing loss. This can be accomplished using genetic counseling of high-risk individuals and families (22, 28). This counseling must be based on the correct diagnosis, which is the responsibility of the audiologist and otologist. Adequate understanding is necessary by the general public and medical profession.

Based on the above-mentioned evidence, consanguineous marriage is an important social health problem that should be addressed by an intensive health education campaign. In the countries such as Iran, where consanguineous marriages are common, there is an urgent need for public education programs and for providing the facilities for genetic counseling and reproductive risk assessment. To increase their awareness of the potential risks of consanguineous marriages, unmarried young females and males, especially those who had a genetic disorder in their families (20), should be targeted by the

educational programs.

Our data demonstrates that 65% of the consanguineous marriages among the parents of deaf children are statistically different than the percentage of consanguineous marriage among Iranian population (38%). This indicates an obvious relationship between severe to profound hearing loss and consanguineous marriage.

Overall there were limitations in our study with regards to the accessibility of parents to interview and data collection. However, our findings showed that hearing loss is more common among consanguineous marriages and their relatives. The effect of a consanguineous marriage is different according to the prevalence of the disease in the family, and it seems that there are methods to prevent hereditary deafness in a population by genetic counseling.

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Footnotes

Authors' Contribution: Dr. Shokofeh Radfar: study concept and design, study supervision, and critical revision of the manuscript; Dr. Mohammad Ajallouyan: analysis and interpretation of data; Dr. Sima Nouhi: scientific co-worker; Dr. Seid Abbas Tavallaie: scientific co-worker; Dr. Susan Amirsalari: scientific co-worker; Dr. Jaleh Yousefi: scientific co-worker; Mahdieh Hassanali Fard: Data gathering.

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