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RE: MS-011-0501 "Impact of Consanguinity on Childhood Hearing Impairment in a Saudi Population"

Dear Dr. Jamal:

Final Acceptance

Thank you very much for submitting the above-titled manuscript. I am pleased to inform you that your *revised* manuscript has been finally accepted for publication in JKAU-Medical Sciences. You are going to receive the actual proof of the above in due course and we will let you know about the date of publication of your article.

I take this opportunity on behalf of the Editorial Board to thank you for your contribution to the publication of JKAU-Medical Sciences.

Yours sincerely,

Editor-in-Chief
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Impact of Consanguinity on Childhood Hearing Impairment in a Saudi Population

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Abstract

Objective:

To determine the current status of consanguinity among Saudi population and its effect on childhood hearing impairment.

Materials and Methods:

A randomly selected sample of 9540 Saudi Children representing all socio-economic and demographic groups were selected. A fieldwork was carried out to enumerate and number the various areas of the provinces of the Kingdom. A survey team of social worker, otolaryngologist, audiologist and a nurse completed the questionnaire form, performed clinical examinations and audiological assessments using free sound speech, tuning fork test, pure tone audiometry using clinical audiometer Acx30 and tympanometry using GSI 33 and Madsen Zodiac 901. The data were analyzed by x2 test using EPI-INFO computer software.

Results:

A total of 9540 children below the age of 15 years were included. Parents of 19% of the children were first cousins and 28% were either second cousins or other relatives. The prevalence of hearing impairment was found to be 13% (0.83% severe, 2.42% moderate and 9.75% mild). The prevalence of hearing impairment was significantly higher in children whose parents were either first cousins (16.14% $P<0.00$) or relatives 12.42% $P<0.01$) as compared to the children whose parents were not related (10.38%).

Conclusions:

Our study clearly demonstrates a high percentage of consanguinity among the Saudi population and a definite role of consanguinity in the etiology of childhood hearing impairment. A well-planned counseling program to create awareness regarding the adverse effects of consanguineous marriage is needed to save the population from the disability of hereditary hearing impairment.

Key Words: Hearing impairment, Genetic factors, Consanguinity, Risk factors

Introduction

Consanguinity (family intermarriages) is commonly practiced in many Asian, African, and Latin America communities. In Saudi Arabia, certain consanguineous marriage as first cousins mating is encouraged as part of the social customs especially among tribes. Certain degrees of kindred like uncle-niece mating are prohibited by religion. The siblings of consanguineous marriage have significantly high incidence of hereditary diseases including hearing impairment.

Hereditary hearing impairment may be conductive or sensorineural (SNHL), most of it is caused by single autosomal recessive gene. Previous study conducted in the city of Riyadh showed high incidence of consanguineous marriage and high prevalence of hereditary sensorineural hearing loss (1,2).

This study was conducted to determine the current pattern of consanguineous marriage in the whole Kingdom of Saudi Arabia, and its impact on childhood hearing impairment particularly on SNHL.

Materials and Methods

A survey of 9540 Saudi infants, pre-school and school age children, below 15 years was carried out during September 1997 to May 2000. The sample selection was randomly designed with representation of children covering all socio-economic and demographic groups from the different provinces of Saudi Arabia. Each survey team was comprised of an ENT specialist, a nurse, a social worker, a field supervisor and a driver. Each child was carefully examined for hearing status and a questionnaire was completed with the help of parents. The questionnaire was based on WHO/PDH ear examination form with some modifications. It consisted of information regarding age, sex, and consanguinity of parents, family history of deafness, hearing and speech deficits and exposure to various known risk factors for hearing impairment.

The hearing impairment in children was tested using free field speech testing and tuning fork tests. Pure tone audiometry (0.5-2 kHs2) and tympanometry were used for further confirmation and the assessment of the severity of hearing loss. Evoked response audiometry was done for the suspected very young children below 4 years of age. The severity of hearing impairment was classified as mild (20-40 dB), moderate (41-70 dB) and severe (71-100 dB) hearing loss. The data were analyzed by X^2 test using EPI-INFO computer software. A value of $P<0.05$ was considered as statistically significant.

Results

Subjects

A total of 9540 Saudi children were included in the study. There were 4189 (43.91%) male and 5351 (56.09%) female children. The age wise distribution of the children was as follows: up to 4 years (2054), >4 to 8 years (3431), >8 to 12 years (3615) and >12 to 15 years (440).

Consanguinity of Parents

The parents of 1809 (19%) children were first cousins, whereas the parents of 2672 (28%) children were either second cousins or other relatives. In case of 4439 (47%) children, the parents had no earlier family relationship, while 620 (6%) subjects failed to give a definite answer (Fig. 1).

Hearing Impairment

Out of 9540 children survey, 1241 (13%) were found to be hearing impaired. The hearing impairment was found to be severe in 79 (0.83%), moderate in 231 (2.42%) and of mild category in 931 (9.75%) children (Fig. 2). The prevalence of hearing impairment was found to be significantly higher in the children whose parents were either first cousins (16.14%, $P<0.001$) or relatives (12.42%, $P<0.01$) as compared to the children whose parents were not related (10.38%) (Fig. 3). The prevalence of SNHL was found to be 1.5%, 10.4% with conductive hearing loss and 1.1% with mixed hearing loss.

Risk Factors

The frequency of some important risk factors in the children with different consanguinity of parents is given in (Fig. 4). The parents who were first cousins showed comparatively high frequency of hearing impairment (5.69%) than the parents who were relatives (3.40%), while it was least in the parents who were not related (1.66%). The past history of hearing impairment was also higher in the families of children whose parents were first cousins (3.59%) followed by its frequency in the families of the children whose parents were relatives (2.05%) or not related (1.68%). The exposure to antenatal (antenatal care not attended) and postnatal (incomplete vaccination) risk factors was quite higher in the children whose parents were first cousins (Fig. 4). However, other risk factors including abnormal pregnancy, abnormal labor, premature birth and low birth weight had no correlation with the consanguinity. The frequency of speech abnormality was quite high in the children whose parents were first cousins (2.65%) as compared to its frequency in children whose parents were either relatives (0.89%) or not related (0.58%).

Discussion

Hearing impairment is a serious disabling condition, especially in the childhood population. Even a mild degree of hearing impairment can result in long lasting communication, social and academic deficit (3). The children with hearing impairment also have poor self-perceptions than the children of same age with normal hearing (4). The prevalence of childhood hearing impairment varies widely in the children from different populations. Naeem and Newton (5) reported a three-fold high prevalence of sensorineural hearing loss (SNHL) in Asian children as compared to non-Asian children. Recent screening of Jamaican children showed the prevalence of SNHL to be 4.9% (6), whereas it was quite higher 8.7% in the children from Tanzania (7), and 11.9% in South India (8). On the other hand the prevalence of hearing impairment has been reported to be comparatively less (0.54 to 2.0 per 1000) in the children from European countries (9,10).

This study showed that consanguineous marriages are quite common in Saudi Arabia (Fig. 1). The parents of 19% children were first cousins and 28% were either second cousins or other relatives. Earlier investigations have also reported high frequency of consanguineous marriages in Saudi Arabia (1,2). We observed an overall prevalence of hearing impairment in Saudi children to be 13% (Fig. 2), which is comparatively higher than that reported in

earlier studies from Saudi Arabia (11). The sensorineural hearing loss was found among 1.5%. There was significantly high prevalence of hearing impairment in the children of consanguineous parents (Fig. 3). A close association between consanguinity of parents and the incidence of hearing impairment in the children has been reported earlier.

The major hereditary deafness has been linked to a single recessive gene. Consanguineous marriages increase the chance of recessive gene inheritance to a greater extent. Bergstrom et al (12) have observed that the children of deaf parent without consanguinity, have only slightly increases risk of hearing impairment due to the rare possibility that both father and mother would affected by the same genetic deafness. On the other hand the children of consanguineous parents are at significantly high risk of hearing impairment, because their parents are more likely to homozygous and capable of passing the trait to their offspring. The effect of consanguinity on the development of childhood hearing impairment also depends on the closeness of the relationship of parents (13). A marriage between first cousins poses a greater risk, whereas a distant consanguinity has comparatively low risk of producing defective offspring, which is also supported by our findings (Fig. 3).

Our results also showed a close association between consanguinity of parents and higher incidence of certain important risk factors including history of hearing impairment in parents and other family members, and speech abnormality in the children (Fig. 4).

First cousins have $1/8^{th}$ of their genes in common and on the average, $1/6^{th}$ of their children would be homozygous for some gene transmitted from a common great-grandparent. Double first cousin marriage has a correspondingly lower risk of producing defective offspring is near enough to the general population risk to be of little or no genetic consequence.

In conclusion, this study clearly demonstrates a definite role of consanguinity in the prevalence of childhood hearing impairment. A well planned counseling program to create awareness about the adverse effects of consanguineous marriages will be helpful to save our population from disability of hereditary deafness. Early intervention for the children affected in the form of hearing aid fitting, speech training and supporting their families during pre-school period and at school are of great value. Those who need cochlear implant should be recommended for the authority. Hearing and speech center should be available at least in big cities to look after these children.

Acknowledgements

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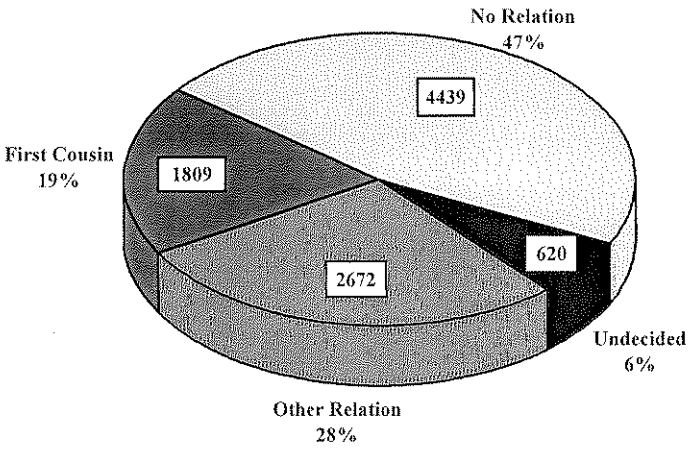

Fig. 1. Frequency distribution of total subjects according to consanguinity of parents.

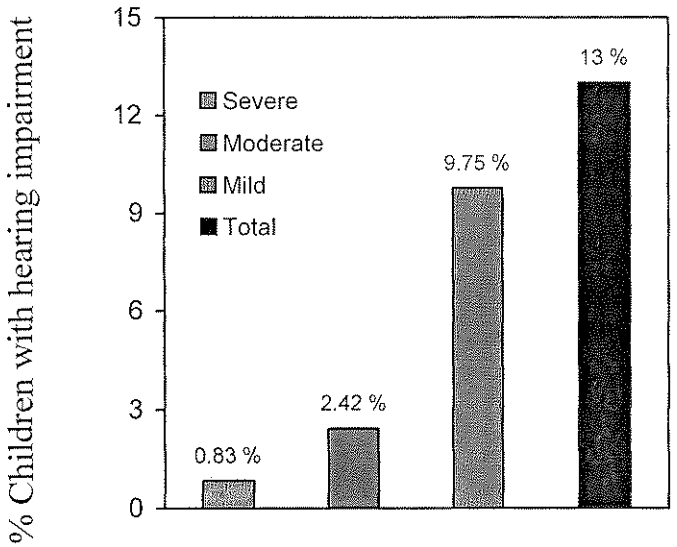

Fig. 2. Prevalence of hearing impairment in the children. Bar graph shows a comparative view of severe, moderate and mild degree of hearing impairment as well as the total prevalence.

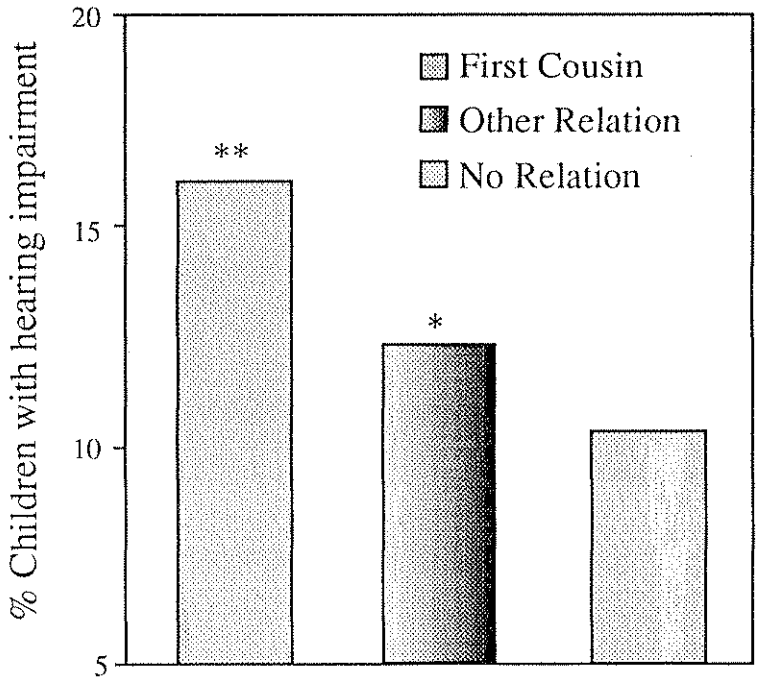

Fig. 3. Effect of consanguinity of parents on hearing impairment in the children. *P<0.01 and **P<0.001 as compared to the prevalence of hearing impairment in the children whose parents were not consanguineous.

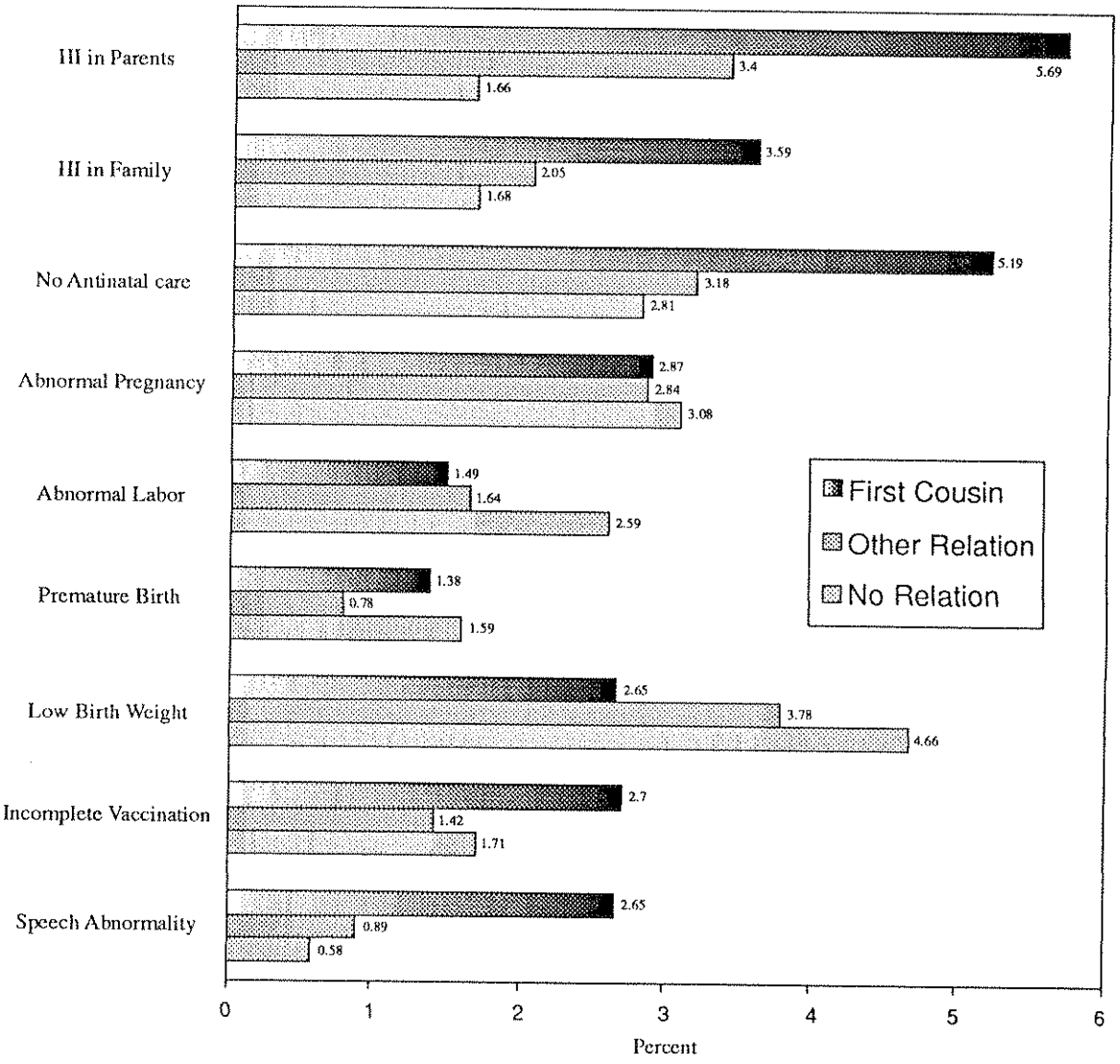

Fig. 4. Frequency of various risk Factors in the Children with different consanguinity of parents. HI = Hearing Impairment.

تأثير زواج الاقارب على ضعف السمع في اطفال السعودية

طارق صالح جمال
قسم الانف و الاذن و الحنجرة، كلية الطب، جامعة الملك عبد العزيز، جدة، السعودية

الملخص: هدف هذا البحث هو معرفة نسبة زواج الاقارب في الوقت الحالي و تاثيرها على ضعف السمع لدى الاطفال. و تكونت الدراسة من اخذ عينة عشوائية بعدد ٩٥٤٠ طفل سعودي تحت سن ١٥ سنة تمثل كل الطبقات و المناطق المختلفة بالمملكة.

ووجد ان نسبة الضعف بين الاطفال الذين لوالدين ابناء عمومة أو اقارب أكبر من نسبتها بين الاطفال الذين لوالدين لا تربطهم قرابة.