

Association of congenital hearing impairment and consanguinity

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Abstract

Background: Consanguineous marriages preferred by many have a major role to play in many congenital problems like hearing impairment. So the objective of the study is to find the Association of Congenital Hearing Impairment and Consanguinity and to know the percentage occurrence of hearing impairment in second and third degree consanguineous marriages. **Methodology:** This Study was conducted in JJM Medical College and Hospital, A total 1356 patients with hearing impairment were selected for the study. Using RMS EMG. EP MARK –II machine latencies of Waves I and V were recorded. Thus, the survey provides information on maternal and child health, fertility, nutrition, contraceptive behaviours, human immunodeficiency virus/acquired immune deficiency syndrome, and other issues. **Results:** This study found that significant number nearly 56% of children with hearing impairment are born to consanguineous marriages, and more than 71% out of these consanguineous marriages are second degree. **Conclusion:** This study concludes that the consanguinity plays a important role in the normal development of hearing so all possible efforts to be done to prevent the consanguineous marriages.

Keywords: Consanguinity, Congenital Hearing Impairment, Wave V.

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INTRODUCTION

Consanguineous marriages are preferred by about one-fifth of the population living in different parts of the world and nearly 8.5% of children are products of these marital unions.¹ Consanguineous marriage is referred to as a marital union between an individual who belongs to the same genealogical kin.² However, consanguineous marriage ranges from cross cousin to more distant relatedness. In countries of North Africa, the Middle East and West Asia where consanguineous marriages are

immensely popular they account for 20%–50% of all marriages.³ Consanguineous marriages are categorized as first, second and third degree, the first being the closest kinship. Second is uncle-niece or first cousin marriages⁴. Third degree is distant blood relatives. First degree considered to be incest, is very rare. First cousin and uncle-niece unions are more frequent, comprising 20%–30% of all marriages. Karnataka (0.3%) witnessed a slight increase in the rates of consanguineous marriages with the uncle, in spite of being the most controversial union.⁵⁻⁶ Man is a social animal and communication is very important for socialization. hearing and speech are the corner stones of communication. Sense of hearing is a gift to mankind from god and speech is snugly related to hearing because those with impaired hearing often have speech delay⁷⁻⁸. Hearing impairment has devastating effects on children as it hampers the child's lingual skills.⁹ Genetic inheritance plays a significant role in children with sensorineural hearing loss, with consanguinity being the chief culprit.¹⁰ Hearing loss in early childhood is of major concern as it leads to poor scholastic performance.¹¹ The most common causes accounting for hereditary hearing loss are

autosomal recessive inheritance followed by autosomal dominant and minor proportion is attributed to X-linked and mitochondrial disorders.¹² Hearing impairment is rapidly increasing sensory deficit amongst human beings and accounts for third of the entire disease burden in the world.¹³ The world wide prevalence of profound, congenital deafness is 11 per 10,000 children, and is attributable to genetic causes in at least 50% of cases.¹⁴⁻¹⁵ The siblings of consanguineous marriages have a significantly higher incidence of autosomal recessive diseases including hearing impairment. Marriages within the family increase the risk of hearing impairment and other diseases. The development of cochlea and hair cells is dependent on a genetic pathway called Planar Cell Polarity (PCP) pathway which is involved in the formation of the polarized structure of the auditory sensory organ and regulates the embryonic development. Genetic disturbances caused due to consanguinity disturb the pathway leading to congenital hearing loss.¹⁶ Consanguineous unions account for 25.8% of all marriages in the southern states of India and with the 28.9% prevalence of consanguineous marriages in Davangere district.⁴ So the present study was done to know the association of congenital hearing impairment and consanguinity in and around Davangere.

METHODOLOGY

This study was conducted in department of Physiology, JJM Medical College, Davangere after obtaining ethical clearance from the institute. 1356 patients under 5 years with hearing impairment were selected for the study. All patients were administered the test procedures with prior appointment. Informed consent was obtained from their parents before the procedure. An ENT check-up was done to rule out the possibility of wax, ear infection, middle ear problems etc. The parents were instructed to wash the scalp of the child thoroughly as a requirement of the test. Prior to the test, each child was examined by the pediatrician and the dosage for sedation was prescribed. Drug used for sedation was syrup Triclofos 20mg/kg body wt. The instrument used was RMS EMG. EP MARK –II machine which is a fully computerized machine manufactured by RMS RECORDERS and MEDICARE SYSTEM Chandigarh. Test was carried out in pre-cooled, quiet, dimly lit room with subject relaxed in supine position with eyes closed. The skin was cleaned with spirit and OMEN abrasive skin preparatory paste. The silver electrode were placed as follows: Cz–vertex, both mastoid, (Ai and Ac) forehead (ground). Resistance was not more than 1ohm. Electrode electrolyte gel was used and electrodes were fixed.

Acoustically shielded THD 32 ear phones were placed on the ear and head bands were adjusted. Monoaural auditory

stimulus consisting of rarefaction clicks of 100 microseconds with intensities starting from 30 dB to 100 dB were delivered through electrically shielded earphones at a rate of 11.1/sec. Contralateral ear was masked. The filter settings used were 150Hz–3000Hz. The polarity used was alternate and the analysis time was 10m/sec. About 2,000 responses were averaged. Latencies of Waves I, III and V and inter peak latency of I-V were recorded. The existence of peak V was considered as sound stimulus heard and perceived by the auditory mechanism. The threshold for each ear was confirmed.

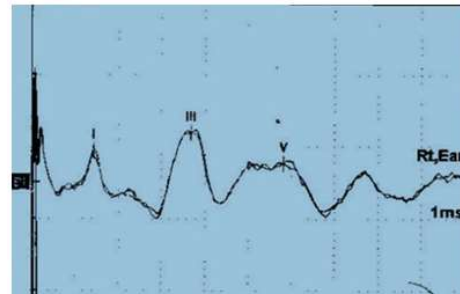


Figure 1: Shows the A normal recording of waveforms.

The guidelines used for the confirmation of peak V were as follows:¹¹

1. Peak V occurs around latency of 5.7 m/sec with S.D. of 0.25 (as per our norms)
2. With decrease, an intensity level latency of peak V increases and its amplitude decreases
3. Peculiar in shape

Normal latency of wave I for infants is 1.47+/-0.19, of wave V is 6.06+/-0.26, and IPL I-V is 4.58+/-0.247

RESULTS

Table 1: The present study found out of these 1356 patients with profound hearing impairment 763 children were born out of consanguineous marriage. 543 were born from second degree consanguineous marriage and the remaining 220 were born from third degree consanguineous marriage.

Table 1: Shows the subjects distribution with percentage

Marriage type	No. of patients	Percentage
consanguineous marriage	763	56.27
Non-consanguineous marriage	593	43.73

Table 2: shows the distribution of the Second degree consanguineous marriages account for 71.17% and the third degree contributed for 28.83% among the subjects.

Marriage type	No. of patients	Percentage
Second degree consanguineous marriage	543	71.17

Third degree consanguineous marriage	220	28.83
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DISCUSSION

Consanguineous marriage is customary in many societies, but leads to an increased birth prevalence of infants with severe recessive disorders. Consanguineous marriage is referred to as a marital union between an individual who belongs to the same genealogical kin¹⁷. However, consanguineous marriage ranges from cross cousin to more distant relatedness, and the rate at which consanguineous marriages occur in a population depends on the population structure and several other factors such as socio demographic, economic, linguistic, geographical characteristics¹⁸. Globally, marriages involving first degree (parents–offspring) and second degree (brother–sister) relationships are called incest and considered to be taboo in almost all the cultures around the world. However, first cousin and uncle–niece unions are more frequent, comprising 20%–30% of all marriages. The most likely reasons for the popularity of such practices can be summarized as a strong family tradition of consanguineous unions, maintenance of family structure and property, strengthening of family ties, financial advantages relating to dowry or bridewealth payments, easy marital arrangements, and a closer relationship between the wife and her in-laws and greater marriage stability and durability¹⁹⁻²⁰. It is therefore often proposed that consanguineous marriage should be discouraged on medical grounds. Panakhian concluded consanguinity is a risk factor for hearing loss. We use this perspective to predict that factors related to agricultural subsistence or traditional intensive kinship systems will increase the likelihood of consanguineous marriage, whereas higher market engagement and associated aspects of family, kinship, and marriage systems will decrease the likelihood of consanguineous marriage. As a result, of consanguineous marriages there is a rise in homozygosis of the pathological recessive genes increasing the risk of birth of handicapped babies. Zakzouk²¹⁻²² reported 50% consanguinity in impaired hearing among 6421 subjects studied from Riyadh city and 9540 from other parts of Saudi Arabia. Al-Gazali carried out a genetic etiological survey of severe childhood deafness in UAE which revealed 74% consanguinity. In another study by Zakzouk *et al.* reported 80.8% consanguinity in their studies carried out on 234 patients from Riyadh with bilateral sensorineural hearing loss that was progressive. This study suggest that the consanguineous marriages cause complications in children should prevent consanguineous marriages.

CONCLUSION

All efforts should be made to prevent consanguineous marriages. The people should be educated about the complications in children born out of consanguinity. Pre-marital counselling has to be done to prevent such marriages.

An objective and non-invasive screening test like BERA need to be done in all the at risk neonates so that abnormality if present can be picked up at an early stage so that, timely early intervention can be done and the normal socio-developmental milestones for the child is not delayed.

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