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Research Article

BERA: A Tool to Study the Impact of Consanguinity on Hearing in Children

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Abstract: Marriage is said to be consanguineous, when it is between persons descending from the common ancestor. Consanguineous marriages are widely contracted worldwide despite the devastating effects in children born to such couples. The aim of the study was to analyze the impact of consanguinity on hearing in babies born out of consanguineous marriages. It was a **c**omparative study in 50 children aged between 1 month -5 years born out of consanguineous marriages and 50 children without parental history of consanguinity who attended pediatric OPD. BERA (Brain stem Evoked Response Audiometry) testing was done with RMS EMG EP MARK-II machine manufactured by the RMS recorders and medicare system, Chandigarh. Students' unpaired t test was used for **s**tatistical analysis. Out of 50 cases of positive history of parental consanguinity, 39 had hearing impairment. No BERA waves were recordable in 17 of 39 children. Remaining 22 had mean wave V threshold 52.36 ± 21.41 dB, which was highly significant statistically when compared to the threshold of 32± 0.1 dB in those without any history of consanguinity. In conclusion, awareness needs to be created in the population to discourage consanguineous marriages. Screening programme should be performed in the nursery and well-baby clinic during immunization in the early neonatal stage to avoid harmful effect on speech and language development.
Keywords: BERA, Consanguinity, Hearing impairment.

INTRODUCTION

Consanguineous marriage is the union of individuals having a common ancestor. It is categorized as first, second and third degree, the first being the closest kinship. It is believed that consanguineous marriages would preserve family dynamics, structure and provide cultural, social and economic benefits. Consanguineous marriages are more prevalent in rural than urban areas and have been positively associated with: Low age at marriage, Low educational level of the mother, Low occupational status of husband.

Sense of hearing is a gift to mankind from god as hearing and speech are the corner stones of communication. Speech is snugly related to hearing because those with impaired hearing often have speech delay [1].

Deafness is usually of two types. It is caused by: (a) Impairment of the cochlea, the auditory nerve, or the central nervous system circuits from the ear, which is usually classified as "nerve deafness." (b) Impairment of the physical structures of the ear that conduct sound itself to the cochlea, which is usually called "conduction deafness. If either the cochlea or the auditory nerve is destroyed, the person becomes permanently deaf. However, if the cochlea and nerve are still intact but the tympanum-ossicular system has been destroyed or "frozen" in place by fibrosis or calcification, sound waves can still be conducted into the cochlea by means of bone conduction from a sound generator applied to the skull over the ear [2].

Hearing impairment has devastating effects on children as it hampers the child's lingual skills [3]. Genetic inheritance plays a major role in children with sensorineural hearing loss, with consanguinity being the chief culprit [4]. Hearing loss in early childhood is of major concern as it leads to poor scholastic performance and ill employment prospects [3]. 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 [5].

MATERIALS AND METHODS

This study was conducted in department of Physiology, JJM Medical College, Davangere during the period from May 2010 to April 2012, after obtaining ethical clearance from the institute. 50 children born out

of consanguineous marriages between 1 month - 5 years from pediatric and ENT out- patient department were taken for the study. 50 age and sex matched children from the general population without history of parental consanguinity formed the control group.

Inclusion Criteria

- All infants and preschool age children at genetic risk, who attend the pediatric OPD of Bapuji Hospital and Chigateri General Hospital, attached to J.J.M Medical College.
- Children with history of parental consanguinity

Exclusion criteria

- Neonatal intensive care of more than 5 days or any of the following regardless of length of stay. Extracorporeal membrane oxygenation (ECMO), assisted ventilation, exposure to ototoxic drugs or loop diuretics (furosemide) and hyperbilirubinemia that requires exchange transfusion.
- In utero infection such as cytomegalovirus, herpes, rubella, syphilis and toxoplasmosis.
- Craniofacial anomalies
- Birth weight < 1500g
- Bacterial meningitis
- Gestational age < 37 weeks
- Apgar scores < 4 at 1 minute or < 6 at 5 minute
- Severe multiple anomalies
- Incompatible with life
- Atresia or stenosis of external ear canal
- Untreated otitis externa

Informed consent was obtained from their parents before the procedure. They were subjected to BERA test. Medicine pedichoryl of dosage 20mg/kg body weight with trade name syrup trichlofos was administered to sedate the child. The area of application of electrodes was cleaned by using abrasive strip. BERA was conducted in a quiet, semi-darkened room. Surface electrodes were placed at the vertex (CZ), both mastoids (Ai and Ac) and forehead (ground). Care was taken to keep the resistance below 5K. Monaural auditory stimulus consisting of rarefaction clicks of 100 microseconds were delivered through electrically shielded earphones at the rate of 11.1/sec. Pure white noise of 40dB was used to shield the contra lateral ear. Responses to 2000 click presentations were averaged [13].

RESULTS

Out of 50 cases with positive history of parental consanguinity, 39 had hearing impairment. No BERA waves were recordable in 17 of 39 children. Remaining 22 had mean wave V threshold 52.36 ± 21.41 dB, which was highly significant statistically when compared to the controls (32± 0.1 dB)without any history of consanguinity. Absolute latencies (in ms) of the wave V were 6.57 ± 0.51 and 6.43 ± 0.35 and IPL III – V were 2.72 ± 0.49 and 2.12 ± 0.32 in consanguineous and non-consanguineous group respectively and the difference was statistically significant. Absolute latencies of Wave I, II, III, IV and IPL of I –III, I – V and amplitude V/I ratio were increased in consanguineous group compared to the non-consanguineous but was not significant.

Table 1: Characteristics of the study group

Gender	No. of cases	Percentage	No. of controls	Percentage
Male	32	64	38	76
Female	18	36	12	24
Total	50	100	50	100

Table 2: Comparison of BERA parameters in children with consanguinity and those without consanguinity

	Consanguinity absent (N= 50)	Consanguinity present (N= 50)	Consanguinity v/s Non Consanguinity	
	Mean±SD	Mean± SD	T value	p value
V (dB)	32.00± 0.10	52.36±21.41	-5.02	<0.0001**
I	1.58± 0.20	1.60±0.22	-0.50	0.62
II	2.77± 0.28	2.92±0.27	-0.63	0.53
III	4.24± 0.26	4.32±0.34	-0.84	0.4
IV	5.47± 0.48	5.32±0.53	1.07	0.29
V	6.43±0.35	6.57±0.51	-2.10	<0.05*
I-III	2.67±0.27	2.54±0.36	-1.10	0.42
I-V	4.77±0.35	4.89±0.42	-1.19	0.24
III-V	2.12±0.32	2.72±0.49	-2.82	<0.05*
V/I	3.50±5.01	5.76±6.24	-1.37	0.18

Unpaired t test, * Significant, ** Highly significant

DISCUSSION

Consanguinity is commonly practiced in all the communities worldwide without paying heed to the detrimental outcome in children. It is an easily preventable cause of genetic disorders which can be executed with increase in awareness and understanding of the problem. It increases the risk of not only hearing impairment but also other kinds of genetic disorders like sickle cell disease, amino acid disorders etc. Such type of marriages results in increased homozygosis of abnormal recessive genes ultimately giving rise to birth of handicapped babies [6].

Consanguinity is a deep seated trend in the society with one billion people currently living in countries where consanguineous marriages are customary, among them; one in every three marriages is between cousins. Uncle-niece marriages are also quite common in our country especially in southern states like Andhra Pradesh, Tamilnadu and Karnataka. The rising public awareness on possible preventive measures for congenital disorders has led to an augmentation in the number of couples seeking preconception and premarital counseling on consanguinity [7].

Similar findings were reported by Reddy MVV *et al*., stating significant association between consanguinity and hearing impairment, thus indicating that the consanguinity is a risk factor for hearing loss [3].

In a study by Razi MS *et al*., the hearing impaired group showed 61% consanguinity as compared to 36% in general population [10]. The prevalence of sensorineural deafness in Bangladeshi children in UK population, was at least 2.3 times the national average, because they were more consanguineous marriages [10].

These results are similar to a study published by Zakzouk, which was conducted on 168 children with sensorineural hearing loss (SNHL), and revealed that consanguineous marriage was responsible for (66.1%) of affected cases. Wave V threshold, absolute latency of wave V (6.58 ± 0.51ms), IPL III-V (2.42 ± 0.49 ms) increased in the cases with history of consanguinity [12].

The effect of consanguinity on the development of childhood hearing impairment depends 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. Hearing impairment in a toddler is of major concern as it hinders with the normal speech and the overall development of the child. In the absence of normal speech, child's ability to communicate is restricted and this has a negative impact on child's social, emotional, cognitive and academic development [8].

CONCLUSION

Awareness needs to be created in the society about the ill effects in off springs born to consanguineous parents. All couples should ideally undergo premarital counseling. Every effort should be made to prevent consanguineous marriages. The rural and illiterate people should especially be educated about the detrimental consequences in children born out of consanguinity as these groups of people favor such marriages on social grounds and to keep the dignity of their spoken words. Non invasive screening tests like BERA need to be done in all the at risk neonates so that we can pick up any abnormality if present, at an early stage so that if possible, timely intervention can be undertaken.

Primary health care providers can counsel for consanguinity provided they possess the recommended education and training. Education of the public in general and of primary health personnel in particular as well as Anganwadi workers plays an important pillar in clarifying the health and social effects of consanguineous marriages [11].

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