© Kamla-Raj 2006 Int J Hum Genet, 6(4): 357-358 (2006)

Role of Consanguinity in Congenital Neurosensory Deafness

M.V.V.Reddy*, L.Hema Bindu, P.P.Reddy** and P.Usha Rani****

Department of ENT, Osmania Medical College, Koti, Hyderabad, Andhra Pradesh, India
Mobile: +91-98490-31935
****Institute of Genetics and Hospital for Genetic Diseases (Osmania University), Ameerpet, Hyderabad 500 016, Andhra Pradesh, India***
Telephone: +91-040-23403681

KEYWORDS Consanguinity; sensorineuro; congenital; hearing loss; syndromic

ABSTRACT Congenital deafness affects the speech and psychosocial development of the affected child. It could be syndromic or non-syndromic depending on the presence or absence of the associated manifestations. Among various etiological factors described for deafness, consanguinity is an established high risk etiological factor. A prospective study was carried out in 1076 children in the age group of 0-14years attending Government ENT hospital and schools for deaf in and around Hyderabad. The results showed that 41.73% (449) of the cases were the products of consanguineous matings and 58.27% (627) were born to non consanguineous parents. Further analysis revealed a high rate of consanguinity (44.53%) in children with non syndromic deafness.

INTRODUCTION

Hearing impairment has debilitating effects on children as it can retard individual's language acquisition skills and impair the overall development. It is rapidly increasing sensory deficit amongst human beings and accounts for one third of the entire disease burden in the world (ICMR 1983). 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 (Marazita et al.. 1993). The recent survey indicates that 1 out of every 1000 children born in India showed profound hearing loss (Kundu 2000). In developed countries about 60% of the cases with isolated deafness are reported to have a genetic origin (Cohen and Gorlin 1995). Genetic inheritance plays a major role in children with sensorineural hearing loss, with consanguinity being the major cause (Turan and Apaydin 2002). Among hereditary deafness, autosomal recessive inheritance predominates accounting for 80% of the cases followed by autosomal dominant inheritance in about 20% of the cases and X-linked and mitochondrial modes of inheritance in less than 1% of the cases (Morton 1991). Consanguineous marriage is a tradition which is commonly practiced among Asian, African and Latin American communities. 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. This pathway 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.

Author for correspondence: Prof.P.P.Reddy, Institute of Genetics and Hospital for Genetic Diseases, Ameerpet, Hyderabad 500 016, Andhra Pradesh, India
Telephone: +91-040-23403681 (off) / +91-040-23113092 (res) / +91-0-9392434664 (mob)
E-mail: reddypenagaluru@rediffmail.com

METHODOLOGY

Epidemiological study was carried out in 1076 children in the age group of 0-14years attending Govt ENT hospital and schools for deaf children in and around Hyderabad. Various audiometric tests such as Pure Tone audiometry (PTA), Oto Acoustic Emissions (OAE) and Brainstem Evoked Response Audiometry (BERA) were carried out on children to know the type and degree of hearing loss. After hearing loss was confirmed, details of family history and consanguinity was recorded. The general population was studied for various parameters including consanguinity. The study was approved by the institutional ethical committee and the parents or guardians of the patients were informed about the study and their consent was obtained.

RESULTS AND DISCUSSION

Out of 1076 children with congenital hearing impairment, 41.73% (449) of the cases were the products of consanguineous marriages and 58.27% (627) of the cases were born to non consanguineous parents (Table 1).

Table 1: Type of marriage of parents of children with hearing impairment

Type of marriage	*No. of cases*	*%*
Consanguineous	449	41.72
Non consanguineous	627	58.27

In the general population consanguinity was 22.36%, where as significantly high percentage of consanguinity was observed among the parents of children with hearing impairment, thus indicating that the consanguinity is a risk factor for hearing loss.

Consanguineous marriages promote a rise in homozygosis of the pathological recessive genes increasing the risk of birth of handicapped babies (Panakhian, 2005). Zakzouk (2002) reported 50% consanguinity in hearing loss among 6421 subjects studied from Riyadh city and 9540 from other parts of Saudi Arabia. A genetic etiological survey of severe childhood deafness in UAE carried out by Al-Gazali (1998) revealed 74% consanguinity. Zakzouk et al. (1995) reported 80.8% consanguinity in their studies carried out on 234 patients from Riyadh with progressive bilateral sensorineural hearing loss. Extensive studies carried out in India from Andhra Pradesh, Tamil Nadu and Karnataka have shown high percentage of consanguineous marriages leading to high percentage of children with amino acid disorders, congenital anomalies and genetic diseases. In the present study the rate of consanguinity was almost double when compared to the rate of consanguinity in the general population. The observations made from the study are shown in table 2. Public awareness about the risks involved in consanguineous marriages though press and electronic media should be undertaken to avoid the birth of children with handicap. Besides the high risk families are advised to go for genetic counseling. Further children of consanguineous matings should be regularly monitored for genetic abnormalities.

Table 2: Rate of consanguinity in general population and in families of the hearing impaired

Type of marriage	*General population*		*Children with HI*	
	No.	*%*	*No.*	*%*
Consanguineous	1297	22.36	449	41.73
Non consanguineous	4503	77.64	627	58.27
Total	5800	100.00	1076	100.00

ACKNOWLEDGEMENT

We are greatful to the superintendent of the Govt. ENT hospital and principals of schools for deaf for their kind cooperation in making this study possible. We are also highly indebted to the parents and the guardians of the patients for their support, information and valuable time given to us.

REFERENCES

Al-Ghazali LI 1998. A genetic aetiological survey of severe childhood deafness in the United Arab Emirates. *J Trop Pediatr,* **44**: 157-160.

Cohen MM Jr, Gorlin RJ 1995. Epidemiological, etiology and genetic patterns. In: RJ Gorlin, HV Toriello, MM Cohen Jr (Eds.): *Hereditary Hearing Loss and its Syndromes.* New York: Oxford University Press pp. 9-21.

Indian Council of Medical Research Publication 1983. Colloborative study on prevalence and Aetiology of Hearing Impairment. *ICMR and Department of Science and Technology,* New Delhi.

Kundu 2000. Status of disability of India. *Rehabilitation Council of India Publication,* p172-185.

Marazita ML, Ploughman LM, Rawlings B, Remington E, Arnos KS, Nance WE 1993. Genetic epidemiological studies of early onset deafness in the U.S. school age population. *Am J Med Genet,* **46**: 486-491.

Morton NE 1991. Genetic epidemiology of hearing impairment. *Ann NY Acad Sci,* **630**: 16-31.

Panakhian VM 2005. Marriage of blood relatives and congenital deafness. *Vest Otorinolaringol*, **2**: 22-24.

Turan O and Apaydin F 2002. Genetic sensorineural hearing loss in childhood. *Kulak Burun Bogaz Ihtis Derg,* **21**: 99-105.

Zakzouk SM, Fadle KA, al Anazy FM 1995. Familial hereditary progressive SNHL among Saudi population. *Int J Pediatr Otorhinolaryngol*, **32**: 247-255.

Zakzouk S 2002. Consanguinity and Hearing Impairment in developed countries: a custom to be discouraged. *J Laryngol Otol*, **116**: 811-816.