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Main Article

Assessment of deafmute patients: a study of ten years

Mangal Singh · S. C. Gupta · Alok Singla

Abstract Present prospective study was conducted on 350 patients presenting with chief complaints of impaired hearing and delayed speech from 1996–2006. The aim of the present study was to find out the prevalence of deafmutism in our area, the aetiology of childhood deafness and to ascertain the role of acoustic reflex test (ART) for hearing screening considering brain stem evoked response audiometry (BSERA) as gold standard. A detailed history, clinical and other relevant systemic examination and investigations were done to find out the cause. All patients were subjected to ART test and BSERA. Male to female ratio was 2.1:1. Prevalence was found to be 5.59 per one lac population in our district. The commonest age of presentation was in the second decade. The causes for childhood deafness were genetic (15.8%), embryopathies (10%), perinatal (10.8%) and postnatal problems (12.5%). In 50.6% cases it was aediopathic. Congenital syndromic abnormalities were found in 5.4% patients. In BSERA 21% patients were found to have residual hearing varying from 60 dB to 100dB. The positive predictive value of ART was found to be low (10.4%). Hence it was recommended that ART should not be used as screening tool for childhood deafness.

Keywords Deafmutism · Prevalence and aetiology · Screening tools

M. Singh · S. C. Gupta · A. Singla
Department of E.N.T. and Head & Neck Surgery,
M.L.N. Medical College,
Allahabad, U.P.,
India

M. Singh (✉)
E-mail: drmangalsingh@rediffmail.com

Introduction

Hearing is essential to learn language and speech and to develop cognitive skills According to WHO, World-wide approximately 350 million people have hearing disorder. The overall prevalence of congenital hearing disorder is 1-3/1000 newborns. The prevalence in high risk group is ten times higher [1]. Glusac, a croatian poet said "I am like a tree-trunk in a mountain deaf, and I might add dumb because I can not speak for not hearing what is spoken." It thematise the anguish, the spiritual pain and increasing loneliness of a man who has lost this vital sense. The neurological development of hearing abilities requires an acoustic stimulation in the first two year of life. Deficits due to absent acoustic stimulation in the first two year of life are nearly impossible to improve by later rehabilitation [2]. If congenital hearing disorders are detected and treated in time, most of the children can develop near normal speech and no special education is necessary [2–4]. For the detection of hearing disorders behavioural observation audiometry (BOA), brain stem evoked response audiometry (BSERA), transient evoked otoacoustic emissions (TEOAS), and acoustic reflex test (ART) with acceptable sensitivity and specificity are available. Early intervention is now possible with the advent of BSERA, which is capable of objectively and accurately screening and confirming hearing loss even in neonates. The aims of the present study were to see the prevalence of deafmutism in and around our district, to find out the possible aetiology and to see the reliability of ART for screening for deafness in children considering BSERA as gold standard. An endeavour was made to find out the residual hearing and suggest remedial measures.

Material and methods

A prospective study was conducted from 1996–2006, which included 350 patients presenting with chief complaints of diminished hearing and delayed speech. Presenting

complaints were noted and relevant family, prenatal, perinatal and postnatal history was asked. Complete ear, nose and throat and whole body examination was done in every patient and his level of hearing was estimated by audiological tests like BSERA and ART on the same date. Percentages were used for nominal variables and comparisons.

BSERA: The machine used for BSERA was Italy made Amplaid MK22. Sedation used was 0.1 ml/kg body weight of inj. Paraldehyde given by intramuscular route. Test parameters were click stimulus (average 2000), morphology of different waves and latency intensity function. The action potential generated in auditory pathway of brainstem recorded five waves. These waves disappear at the level of threshold. Wave I represent cochlear nerve, II cochlear nucleus, III superior olivary nucleus. IV lateral lemniscus and V inferior colliculus [5, 6]. At intensity where wave V disappeared +/– 10db was taken as hearing threshold. Two waves at the same intensity were recorded and overlapped for seeing consistency of different waves.

ART: The instrument used for ART was Seimen's SD-30 Impedance Audiometer. For the measurement of acoustic reflex threshold, hand held probe tip assembly was sealed in the external auditory canal of the patient. For screening pure tone stimulus of 90 db was given at different frequencies of 500, 1K, 2K and 4 K Hz and reflex pattern was observed.

Results

The male female ratio was 2.1:1. The commonest age of presentation was second decade constituting 45.4% (Fig. 1). Out of 350 patients, 79% were from Allahabad district and rest 21% were from surrounding districts like Mirzapur (10%), Sultanpur and Kaushambi 4% each and 3% from Jaunpur. The prevalence of deafmute patients in Allahabad district was found to be 5.59 per 100000 of general population during last 10 years when population of Allahabad district was 49,36105 (2001 census).

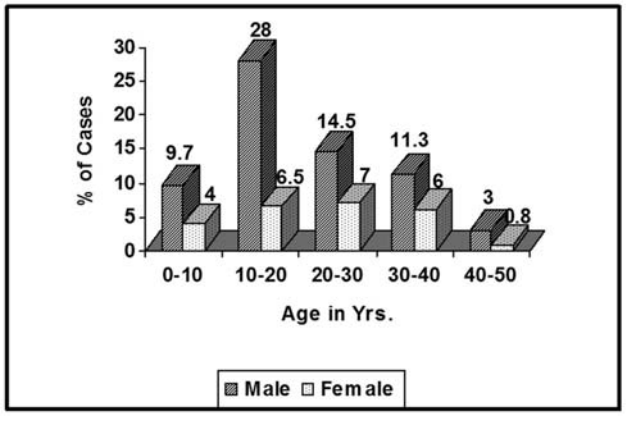

Fig. 1 Age and sex ratio in 350 patients

The probable cause of deafness as per history and clinical examination has been illustrated in Table 1. Out of nineteen syndromic abnormalities, three cases of usher syndrome, four waardenberg syndrome, two down syndrome and one patient of treacher collin syndrome were found. Seven patients had associated nonspecific ear deformities and five had cardiac anomalies. Figure 2 shows a family of waardenberg syndrome with five affected siblings. Three were having characteristic features of hearing impairment, vitiligo, white forelock, dystopia canthorum, heterochromia irides and nystagmus.One patient of treacher collin syndrome presented with microtia, external auditory canal stenosis, cleft palate, malar hypoplasia with slanting palpebral fissures.

In the present study, ART was done in 250 deafmute patients. Out of 250 patients, acoustic reflex was present in 75 patients (30%) at 90 dB of pure tone stimulus. Ten percent of these patients had reflex present in both ears, 10.8% right ear and 9.2% left ear only. BSERA was done in 120 patients,

Table 1 Etiological factors for deafness

Sl. no.	Etiology	No. of patients
	Non-genetic causes	33.3%
1.	Embryopathies	35(10%)
	(a) Infection	12
	(b) Toxaemia of pregnancy	8
	(c) First trimester bleeding	4
	(d) Ototoxic drugs	5
	(e) Jaundice	4
	(f) Rh incompatibility	2
2.	Perinatal causes	38(10.8%)
	(a) Low apgar score	16
	(b) Low birth weight (<2.5 kg)\ prematurity	12
	(c) Breech presentation	5
	(d) Post-term	2
3.	Post-natal causes	44(12. 5%)
	(a) Eruptive fever	12
	(b) Meningitis	8
	(c) Hyperbilirubinemia	5
	(d) Traumatic	4
	(e) Cerebral palsy	5
	(f) Delayed mi1estones	10
	Genetic causes	56(15.8%)
1.	Family history	38(10.8%)
	(a) Paternal	16
	(b) Maternal	12
	(c) Siblings	10
2.	Congenital syndromes	19(5.4%)
	Idiopathic	177(50.6%)

79% showed no response at 100 db threshold where as 21% had residual hearing at different threshold of stimulus as illustrated in Fig. 3. We compared BSERA and ART results of 100 deafmute patients to find out the positive predictive value of ART i.e. proportion of patients with absent acoustic reflex who actually had deafness as observed by BSERA.

Fig. 2 A family of waardenberg syndrome

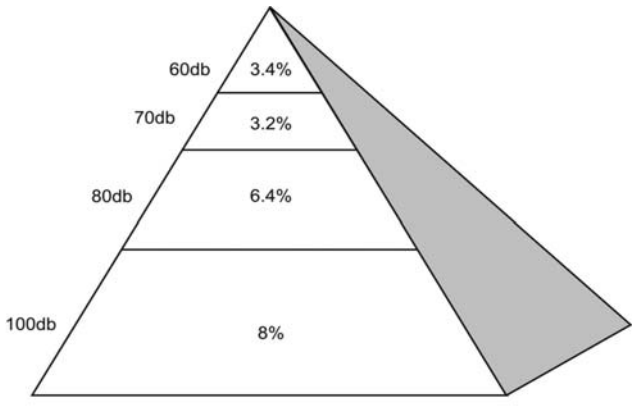

Fig. 3 Residual hearing in deafmute patients as observed by BSERA

It was observed that residual hearing was actually present in only 3.2% of 30 patients with positive acoustic reflex at 90db threshold. Hence, the positive predictive value of ART was found to be-very low 10.4% (Table 2).

Discussion

There is wide variation across the globe in the incidence and prevalence of childhood hearing loss and its possible etiology. Prevalence was higher in Jamaican children (4.9%) [7], Tanzania and South Africa (8.9 to 11.9%) [8], Saudi Arabia (13%), Australia (16/1000) [9]. The incidence per thousand live birth was 1/1000 in USA to 2 to 3.5 in Europe [10–12]. In India according to 1981 census, 3 lacs deafmute children were there and two deaf babies are born per hour. Thus, 18000 deaf babies are added to our population every year [13]. Similarly, there is wide variation in the aetiology of childhood deafness in different parts of the world. Fraser [14] in (1960) from U.K. reported that in 70% patients, the aetiology was congenital and prenatal problem and in 30% it was acquired causes. Strauss [15] in 1990 from USA reported that the probable causes of congenital deafness in their patients were toxoplasmosis (10–15%), rubella (33%) aid cytomegalo virus (33–48%). He further observed that with introduction of immunization programme, the incidence of the disease has decreased. The most frequent cause of acquired deafness in childhood was meningitis [16, 17]. The incidence of post-meningitic deafness varies from 3.5 to as high as 37.2% [l8]. Preterm babies have higher incidence of hearing loss than normal because of prolonged hypoxia or acidosis [19].

The diagnosis of deafness in children had always been a challenge. It is difficult to establish threshold by most commonly used subjective methods like behavioural observation audiometry (BOA). The currently available objective methods for hearing screening in infants are TEOAE and ART in impedence audiometry and finally the BSERA for establishing the threshold of hearing. The TEOAEs machine is not yet widely available in India. However, the impedance audiometer is widely available. Hence this study was planned to find out the prevalence and aetiological profile of childhood deafness in this part of the country and. further to see the reliability of ART for hearing screening considering BSERA as gold standard.

Table 2 Comparison of ART and BSERA results in 100 patients

Types of audiological Test	Reflex absent	Reflex present
ART	70%	30%
BSERA (no response group; 79%)	52.2%	26.8%
BSERA (Residual hearing present group; 21%)	17.8%	3.2%

The prevalence of childhood deafness was found to be 5.59 per one lac of general population. This figure is lower as compared to international statistics. The lower figures in the present study could be due to the fact that it was not a population-based survey, only a hospital based statistics. Lot of parent might not have reported their deaf children to hospital in the city. In 50% of our cases, etiology could not be ascertained. Genetic and post-natal problem were the cause in about one fourth of cases. It was obvious from our study that most of the causes of deafness are preventable by improving the standard of obstetric care by increasing the general socio-economic status. However, it was a distressing finding that most of our patients presented in 2nd decade of life when learning ability diminishes and rehabilitation is difficult. This trend should be reversed by increasing mass awareness and doing hearing screening programmes of at risk babies in their 1st year of life.

In the present study, acoustic reflex was present in 26.8% of patients in spite of fact that no residual hearing was present in them. Out of 21% of patients with residual hearing at 60 to 100 db of threshold confirmed by BSERA, only 3.2% had acoustic reflex present at 90 db. Hence, considering BSERA as gold standard, positive predictive value of ART was found to be very low (10,4%). This confirmed that ART is not a reliable screening test in childhood deafness. According to Bellman and Vanniasegaram [20], the presence of an acoustic reflex may be useful in non-organic hearing loss but it would be unwise to rely only on this measurement. In the presence of a cochlear hearing loss, there is 90% chance of observing a reflex if the hearing threshold is less than 60 db. This was assumed to be related recruitment of loudness in cochlear hearing loss. However, if the acoustic reflex was absent with a sensorineural hearing loss of 60 db, it was more likely to be a retrocochlear hearing loss [21]. Further Niemeyer and Sesterhenn [22] observed that as the severity of hearing loss increased the difference between the pure tone and noise induced reflexes decreased. The method is clearly not as accurate as the objective test using electric response audiometry recordings.

Conclusion

The incidence of deafmute patient in Allahabad zone is low. The aetiology was multifactorial, genetic and postnatal problem being the most common (28% of patients) most of pedigree analysis was paternal in origin. Congenital syndromes constituted 5% only. ART was not found to be reliable screening test for assessing hearing in suspected deaf patients comparing BSERA as gold standard.

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