International Journal of Otorhinolaryngology and Head and Neck Surgery
Shrivastava OP et al. Int J Otorhinolaryngol Head Neck Surg. 2018 Sep;4(5):1193-1197
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

Original Research Article

DOI: http://dx.doi.org/10.18203/issn.2454-5929.ijohns20183444

Demography and etiology of congenital sensorineural hearing loss in children

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Received: 22 July 2018
Accepted: 10 August 2018

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ABSTRACT

Background: The prevalence of sensorineural hearing loss (SNHL) is 2-3 per 1000 live births in India, and out of 1000 babies, 1 is having profound degree of hearing loss at birth or in the pre-lingual age group. Hearing loss is categorized as syndromic or non-syndromic hearing loss. The present study was done to identify and diagnose children with SNHL and study the etiological profile, so that all necessary timely precautions may be taken as SNHL in pediatric age group affects not only development of speech and language, but also emotional, social and behavioural development.
Methods: A cross-sectional observational study was carried out in Department of ENT of a tertiary care centre, in 50 children with congenital SNHL. Data on epidemiological and etiological factors were collected. All cases were analyzed for detailed history and underwent clinical and audiological assessment.
Results: Majority of cases presented between 2 and 4 years of age, however age of detection of congenital deafness by parents was most commonly observed in 6-12 months of age. Out of 50 cases, the maximum number of cases [14 (28%)] were having history of antenatal complications, 11 (22%) cases were having adverse perinatal and postnatal history, while in remaining cases, the etiological factors remained undetermined. In one case associated syndrome was observed (Van der Hoeve's syndrome).
Conclusions: The increased occurrence of etiologies in this study was of prenatal origin, followed by perinatal origin. The present study conducted at a tertiary centre reflects the need of 'High risk' registry maintenance with active surveillance and subsequent screening for early detection of hearing loss.

Keywords: Congenital hearing loss, Sensorineural hearing loss, Etiology

INTRODUCTION

The prevalence of sensorineural hearing loss (SNHL) is 2-3 per 1000 live births in India, and out of 1000 babies, 1 is having profound degree of hearing loss at birth or in the pre-lingual age group.[1]

Hearing loss is categorized as syndromic or non-syndromic hearing loss. Approximately 70% of congenital cases in association with genetic factors are categorized as non-syndromic and the remaining as syndromic deafness because of clinical findings.[2] Non genetic congenital hearing loss is often attributed to prenatal infections with neurotrophic viruses such as rubella, toxoplasmosis, cytomegalovirus (CMV), measles and mumps. A recent study suggested that "more than 40% of deafness of unknown cause, needing rehabilitations for communication" is attributed to CMV.[3]

Most inner ear malformations arise when formation of the membranous labyrinth is interrupted during the first trimester of pregnancy.[4] This interruption may be either a

result of inborn genetic error or a consequence of a teratogenic exposure during the period of inner ear organogenesis between the fourth and eighth week of gestation.

Walch et al in their study to determine the etiology of bilateral sensorineural hearing disorder found that age of first diagnosis was 4 months to 11 years with a mean of 42 months. The sex ratio of male-female was found to be approximately 1:1. The cause of hearing impairment was found to be acquired in 38% of the cases and genetically inherited in 18%. However, in 44% of the cases, the etiology of hearing loss could not be determined.[5] Sutton and Rowe in 1997, in a study to find the risk factor for childhood sensorineural hearing loss found significantly increased number of cases with hearing loss in families with low socio-economic status.[6]

The present study was done to identify and diagnose children with SNHL and study the etiological profile, so that all necessary timely precautions may be taken as SNHL in pediatric age group affects not only development of speech and language, but also emotional, social and behavioural development.

Aims and objectives

To identify and diagnose children with congenital severe to profound SNHL and study their etiological profile.

METHODS

The study was carried out in the Department of ENT of a tertiary care centre during the period of October 2009 to October 2011 on 50 patients having sensorineural hearing loss since birth.

Inclusion criteria: The cases were selected from patients attending Department of ENT for hearing impairment and delayed development of speech and language since birth.

Exclusion criteria: Subjects having normal or near normal level of hearing before developing hearing loss were excluded from study. Subjects having sensory neural hearing loss of less than 70 dB bilaterally were also excluded from study.

Examination of patient includes general physical and systemic examination, ear, nose, throat examination and psychologist consultation.

Clinical audiological evaluations include pure tone audiometry, impedance audiometry, brain stem evoked response audiometry, and oto-acoustic Emission to determine the type and degree of hearing loss.

Data on socio-demography and etiological factors were collected which included detailed medical history of child and mother, socioeconomic status, parent's occupation and literacy, age, sex, religion, birth order, details of developmental milestone and family history of SNHL in siblings. Details regarding parental age at conception, maternal health (diabetes, hypertension, cardiac disease, thyroid dysfunction), reproductive history, history of addiction (smoking, alcohol, gudakhu), exposure to X-ray, maternal infections (fever, rashes), birth history details like birth trauma, birth asphyxia, prematurity, neonatal jaundice, history of acquired factors (postnatal) like meningitis, head injury, ototoxic drugs, infections like measles and mumps were recorded using a standard questionnaire, prepared for this study.

RESULTS

On evaluation of 50 cases presenting with hearing impairment and/or under or delayed development of speech, the age of patients ranged between 1 year and 18 years with mean age of 5.44 years. Maximum patients (36%) were in age group of 2-4 years. Male patients showed higher percentage in age group of 2-4 years (47.83%); while females showed higher percentage in age group of 4-6 years (37.04%). Male:Female ratio observed was 1:1.17 with female preponderance (Figure 1).

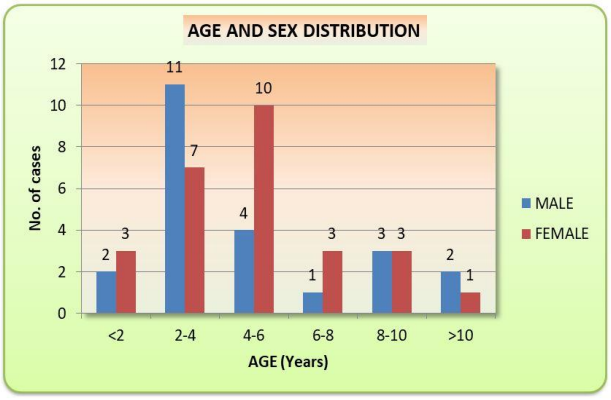

Figure 1: Age and sex distribution.

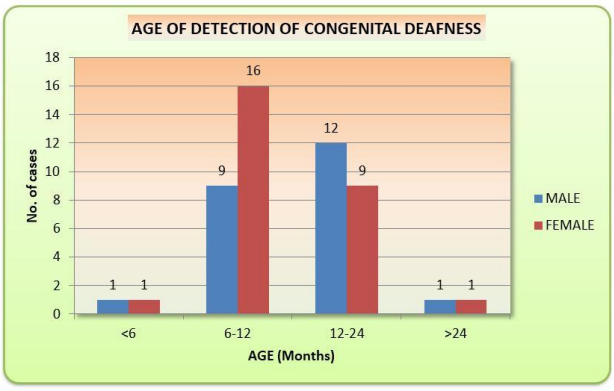

Figure 2: Age of detection of congenital deafness.

The range of age group when the parents suspected the hearing impairment was between 6 months and 30 months with mean age of 15.50 months. Age of detection of congenital deafness by parents was most commonly

observed in 6-12 months of age (50%) followed by 12-24 months age group (42%) (Figure 2).

Table 1: Observation of adverse antenatal history.

S.No.	Antenatal symptoms/causes	Total cases (%)
1.	Fever	1 (7.14)
2.	Rashes	1 (7.14)
3.	Drug intake	2 (14.29)
4.	DM/HT/Cardiac disease	1 (7.14)
5.	Jaundice	1 (7.14)
6.	Addiction	2 (14.29)
7.	Bleeding per vagina	3 (21.43)
8.	Abnormal weight gain	2 (14.29)
9.	Pedal edema	1 (7.14)
	Total	14 (28)

Adverse antenatal history was observed in 28% cases. Out of which, most commonly observed antenatal cause was bleeding per vagina (21.43%) followed by drug intake (anti-hypertensive drug), addiction (gudakhu) and abnormal weight gain (14.29% each) (Table 1).

Table 2: Observation of adverse perinatal and postnatal causes.

S.No.	Perinatal cause	Total cases (%)
1.	Low birth weight	1 (9.09)
2.	Kernicterus	2 (18.18)
3.	ICU Hospitalization	2 (18.18)
4.	Developmental delay	3 (27.27)
5.	Delayed cry	2 (18.18)
6.	Associated syndrome	1 (9.09)
	Total	11 (22)

Adverse intranatal and postnatal history was observed in 22% cases. Out of which, most commonly observed cause was developmental delay (27.27%) followed by kernicterus, ICU hospitalization and delayed cry (18.18% each). In one case (9.09%) associated syndrome was observed (Van der Hoeve's syndrome) (Table 2).

Audiological assessment

On audiological evaluation of 50 cases of congenital deafness, 41 children cooperated for pure tone audiometry. Profound sensorineural hearing loss was observed in 34/41 cases (82.93%) in right ear and 35/41 cases (85.37%) in left ear. Severe sensorineural hearing loss was observed in 7/41 cases (17.07%) in right ear and 6/41 cases (14.63%) in left ear. Otoacoustic emission was absent in 96% cases in bilateral ears and present in remaining 4% cases in bilateral ears. In all 50 cases of congenital deafness, Brainstem evoked response audiometry showed absence of wave V at 90 dB threshold. Psychological examination was normal in all patients.

DISCUSSION

In present study, a total of 50 patients with hearing impairment and/or under or delayed development of speech and language presenting to the ENT outpatient department, of tertiary care centre from October 2009 till October 2011 were observed.

Sex ratio

Bamiou et al observed male preponderance in children with sensorineural hearing loss (59.8%), however no statistically significant sex difference was observed, in terms of severity of hearing loss.[7] Kalsotra et al also observed male preponderance of 1.72:1 in a study of 261 cases of congenital and early acquired hearing loss consisting of 165 males (63.2%) and 96 females (36.8%). However, they observed female preponderance in chromosomal and multi factorial inheritance which has been well documented by Das and Newton.[8]

The present study of 50 cases consisted of 23 males (46%) and 27 females (54%) with female preponderance of 1:1.17. The findings correlate with those of Sanjay et al. (2011) who reported male and female distribution to be approximately equal.[9] However, many earlier studies show male preponderance.[10-12] This may be attributed to the fact that now a day's awareness for female child has been increased as compared to past.

Age of presentation

The age of presentation observed by Kalsotra et al was variable, the range being from less than 1 month to 22 years (mean age=6.73 years). 40.5% of cases presented before 4 years of age with peak at fourth year (16%).[8] Nikolopoulos et al evaluated 108 children before cochlear implantation surgery and children's ages ranged from 21 months to 16 years (mean age, 5.4 years) in their study.[13]

In present study, the age of patients presenting to ENT outpatient department ranged between 1 year and 18 years (mean age=5.44 years, SD=3.43). Maximum patients (36%) were in age group of 2-4 years. Male patients showed higher percentage in age group of 2-4 years (47.83%) followed by 4-6 years (17.39%); while females showed higher percentage in age group of 4-6 years (37.04%) followed by 2-4 years (25.93%). Our study is compatible with the previous study of Nikolopoulos et al and Kalsotra et al.[8,13]

Age of detection

Kalsotra et al observed the range of age group when the parents suspected the hearing impairment was right from birth due to presence of congenital external ear malformations to as late as 16 years (mainly in females of

marriageable age desirous of early treatment). They observed mean age of 2.54 years.[8]

In present study, the range of age group when the parents suspected the hearing impairment was between 6 months and 30 months (mean age=15.50 months, SD=6.48). Age of detection of congenital deafness by parents was most commonly observed in 6-12 months of age (50%) followed by 12-24 months age group (42%). Male patients were detected most commonly in the age group of 12-24 months (52.17%), while females were detected most commonly in the age group of 6-12 months (59.26). In present study, mean age of detection was less as compared to previous study. This discrepancy may be due to increased awareness for hearing impairment now a day.

Etiology of congenital deafness

Antenatal causes

Kalsotra et al observed maternal Rubella as the most common cause of prenatal group of hearing loss. In present study rashes were observed in 7.14% cases during antenatal period, but they were not confirmed as case of rubella.[8] Luiz et al in a study of 104 patients undergoing cochlear implant surgery also observed congenital rubella as the most common etiology of deafness in antenatal period.[14] Most common antenatal symptom in present study was bleeding per vagina.

Perinatal and postnatal causes

Kalsotra et al observed birth anoxia and prematurity as common causes of hearing loss perinatally followed by neonatal jaundice.[8] Luiz et al in a study of 104 patients undergoing cochlear implant surgery observed perinatal hypoxia in 10 % cases and prematurity in 3% cases.[14]

Out of 50 cases of congenital deafness, home delivery was observed in 38 cases (76%). Adverse intranatal and postnatal history was observed in 11 cases (22%). Out of which, most commonly observed cause was developmental delay (27.27%) followed by kernicterus, ICU Hospitalization and delayed cry (18.18% each). In one case (9.09%) associated syndrome was observed (Van der Hoeve's syndrome). In present study the causes we found are mostly due to the lack of general awareness regarding the antenatal care and home delivery.

CONCLUSION

Congenital sensorineural hearing loss is fairly common in children. In present study, mean age of presentation of congenital sensorineural hearing loss to tertiary health centre is 5.44 years and mean age of suspicion or detection of hearing loss by parents is 15.50 months. So, there is a need to educate the people regarding advantage of early intervention and rehabilitation for hearing loss. The increased occurrence of etiologies in this study was of prenatal origin, followed by perinatal origin. The present study conducted at a tertiary centre reflects the need of 'High risk' registry maintenance at all health centres with active surveillance and subsequent screening for early detection of hearing loss. Early education and rehabilitation can then be instituted, allowing these children to reach their potential.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

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Cite this article as: Shrivastava OP, Gupta A. Demography and etiology of congenital sensorineural hearing loss in children. Int J Otorhinolaryngol Head Neck Surg 2018;4:1193-7.