Characteristics of Hearing Impairment in Children Aged Six Months to Two Years with Global Developmental Delay

Ramesh Bhat Y, Harish Kashyap, Pushpa Kini and Shrikiran

Department of Paediatrics, Kasturba Medical College, Manipal, Manipal Academy of Higher Education (MAHE) University, Karnataka, India

ABSTRACT

Introduction: Children with global developmental delay (GDD) are at greater risk to have hearing impairments. These impairments interfere with developmental progress or rehabilitation effects. Hearing impairments may be correctable and if so, may improve developmental outcomes. We aimed to study the incidence, characteristics and probable risk factors of hearing impairment in children aged six months to two years with GDD.

Methods: In this prospective study, an auditory evaluation was carried out by a trained audiologist in children with GDD. Transient evoked otoacoustic emissions (TEOAE) and brainstem auditory evoked response (BERA) were assessed in these children. Hearing loss was classified based on Goodmann's classification.

Results: Of 113 children with GDD assessed, hearing impairment was identified in 35 (30.9%) children. Of 35 children, 22 (62.8%) had isolated sensorineural hearing loss (SNHL). Conductive hearing loss was identified in four (11.5%) and combined hearing loss in nine (25.7%). Of 31 children with SNHL, hearing loss was bilateral in 25 (80.6%). In SNHL, hearing loss was profound in one (3.2%), severe in 14 (45%), moderately severe in three (9.6%), and moderate in five (16.2%). Hearing impairment was associated with 31.7% (20/63) children with cerebral palsy. Kernicterus was another predominant risk factor associated with SNHL. Metabolic disorders, otitis media, tuberous sclerosis, and metachromatic leukodystrophy were other conditions associated with hearing impairment.

Conclusions: Hearing impairment is accompanied by about a third of children with GDD. The hearing impairment in children with GDD tends to severe SNHL type in about 45% and bilateral in about 80%.

Keywords: BERA; Children; GDD; Hearing impairment; SNHL

Correspondence:
Ramesh Bhat Y
Department of Paediatrics
Kasturba Medical College, Manipal,
Manipal Academy of Higher Education (MAHE) University,
Karnataka, India
Email: docrameshbhat@yahoo.co.in

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INTRODUCTION
Significant hearing impairment, if undetected, will impede speech, language and cognitive development in children. Hearing loss in infants may be peripheral or central in origin. It may be unilateral or bilateral; conductive, sensorineural, or mixed; mild, moderate, severe or profound; of sudden or gradual onset; stable, progressive or fluctuating. The effects of hearing impairment depend on the nature and degree of the hearing loss and the individual characteristics of the child. Factors such as intelligence, medical and physical conditions including associated syndrome, family support, age at onset, time of detection, and interventions also affect the impact. About 20% of under-five children with global developmental delay (GDD) were reported to have abnormal brainstem auditory evoked response (BERA) and about 60% of children with cerebral palsy (CP) were found to have sensory neural hearing loss (SNHL). Currently, the average age of detection of the significant hearing loss is reported to be 14 months. Developmental disabilities are estimated to affect 5% to 10% of children. The prevalence of GDD in the paediatric population is not precisely known. The estimated prevalence range from 1% to 3%. GDD has variety of causes. It is associated with decreased adaptation and learning skills. Hence GDD children have problems in activities of daily living. It is likely that children with GDD are at higher risk to have vision and hearing impairments. Evaluation of such impairment is usually a routine practice at the initial management of children with GDD. Hearing impairment interferes with developmental progress or rehabilitation effects. The impairments may be correctable and such correction likely positively influences developmental outcomes. Detection of a specific type of hearing deficit may also help establish the etiology of an underlying developmental disorder.

Kwok et al studied 260 children with severe GDD and found deafness in 18% of children. Rupa et al. in their study involving 96 children with GDD and clinically suspected hearing loss, found hearing loss 91%. Transient evoked otoacoustic emissions (TEOAE) has been found to be a screening tool for hearing impairment in children. In this context, we aimed to assess the incidence of hearing impairment in children aged six months to two years with GDD, study its characteristics, and to identify its risk factors.

METHODS
A prospective study was carried out over 21 months in the Department of Paediatrics, Kasturba Hospital, Manipal, India. The study population included children admitted with GDD in the age group, six months to two years. Children under six months of age or above two years of age and children whose parents were unwilling for hearing assessment were excluded. Approval from the institutional ethics committee was obtained for the study. The purpose of the study was explained to the parents and consent was obtained. Detailed history regarding developmental delay was taken and clinical examination was performed. A pre-structured proforma was used to record the relevant information from individual cases. GDD was based on parental reporting measures, a checklist of developmental milestones from Nelson Textbook of Paediatrics, and clinical examination details by two or more paediatricians. Children were subjected to auditory evaluation by a trained audiologist. TEOAE and BERA were assessed in these children. Results of TEOAE and BERA were recorded and interpreted accordingly. The workup for the etiology of GDD was carried out depending upon the findings from the history and detailed clinical examination. A duly calibrated OAE instrument of ILO DP Echoport 292 Usb II V.6 with foam tip was used to obtain TEOAE measurements. All the evoked potential data were obtained using Intelligent Hearing System (IHS) hardware with software version Smart EP 4.03. Hearing tests were carried out in a sound-treated air-conditioned room with adequate illumination. TEOAEs were obtained for click stimuli presented at a level of 80dB. Stimuli were acquired using non-linear stimulus mode where in stimuli were presented in groups of four. Signal to noise ratio above 6 dB at a minimum of three frequencies was considered as the presence of OAE. ABR recordings were obtained when the child was either in natural sleep or sedated with orally administered Pediclyrol (Triclofos) at a dose of 100 mg/kg body weight. The electrode site was cleaned using a skin
preparation gel. Gold cup electrodes were placed with the help of conduction gel. The placement of electrodes was done according to the International 10-20 classification. All the evoked potential data was obtained using Intelligent Hearing System (IHS) hardware with software version Smart EP 4.03. The stimulus used was click stimulus and was presented using standard insert earphone at 40 dBnHL, EEG was acquired using a single channel (3 electrodes) with horizontal electrode montage (Fz –M1/M2). Auditory brainstem response (ABR) waveforms were analysed for the presence or absence of waves I, III and V as well as their latencies till 30 dBnHL. GDD was defined as significant delay with developmental quotient (DQ) < 70% in ≥ 2 of the major developmental domains (Gross motor, fine motor, language and personal-social). CP was considered when there was a disorder of posture, movement and tone due to static encephalopathy acquired during brain growth in the perinatal period or early infancy. Kernicterus or sequel of bilirubin encephalopathy was defined when a neurologic syndrome resulted from a high level of unconjugated bilirubin in the neonatal period with suggestive MRI findings in the brain. Hypoxic ischaemic encephalopathy was defined as the presence of at least one of the following: not cried at birth and required resuscitation in the form of bag and mask or bag and tube ventilation; persistence of APGAR score 0 - 5 for longer than 5 min; neonatal neurologic sequelae (Seizures / coma / hypotonia) requiring care at the hospital with or without multiple organ involvement. Dysmorphic features are considered when there is a morphologic defect of an organ, part of an organ or larger region of the body resulting from an intrinsically abnormal developmental process. Microcephaly was defined as a head circumference that measures more than three standard deviations below the mean for age and sex. Metabolic disorders as an etiology for GDD was considered in the absence of significant perinatal insults, structural brain malformations, syndromic features or chromosomal abnormalities along with positive results in metabolic screening. Metabolic testing included arterial blood gas, lactate, pyruvate, ammonia levels, creatinine phosphokinase and urine analysis. No etiology for GDD was considered when there was no significant perinatal

| Characteristics | n  | %  |
|-----------------|----|----|
| Age             |    |    |
| 6 mo to 12 mo   | 57 | 50.5 |
| 13 mo to 18 mo  | 33 | 29.2 |
| 19 mo to 24 mo  | 23 | 20.3 |
| Sex             |    |    |
| Male            | 68 | 60.2 |
| Female          | 45 | 39.8 |
| Maturity at birth |   |    |
| Preterm         | 17 | 15  |
| Term            | 96 | 85  |
| History of parental Consanguinity | 22 | 19.5 |
insult, structural brain malformations, or syndromic features and metabolic workup was negative. Hearing loss was graded based on Goodmann classification as follows: normal < 15 dBnHL; slight hearing loss 15 - 25 dBnHL; mild hearing loss 26 - 40 dBnHL; moderate hearing loss 41-55 dBnHL; moderately severe hearing loss 56 - 70 dBnHL; severe hearing loss 71 - 90 dBnHL and profound hearing loss > 90 dBnH.

RESULTS
A total of 113 children with global developmental delay in the age group of six months to two years were admitted and evaluated for hearing impairments. About half of the children (50.5%) were in the age group of six months to 12 months (Table 1). The male to female ratio was 1.5:1. The mean age at presentation to our centre was 13.5 months and the median was 12 months. The history of parental consanguinity was present in 19.5%. Microcephaly was found in 69 children (61.1%) (Table2). Syndromic features were found in three (2.6%) children. All three children were noted to have Down syndrome. Dysmorphic features were found in 40 (35.4%) children and they all had two or more dysmorphic features. Specific dysmorphic features found in the present study included i) Craniofacial (n = 60): low set ears (Nine), brachycephaly (Six), flat occiput (Seven), squint (Seven), epicanthal folds (Six) and craniosynostosis (Seven). High arched palate and sloping forehead were observed in three children each. Malformed pinna, coarse facial features, and telecanthus were observed in two children each. Preauricular tag (One), bulbous nose (One), turricophage (One), retrognathia (One), and synophrys (One) were the other dysmorphic features. ii) Extremities (n = 21): Unilateral simian crease and clinodactyly were seen in five children each. Bilateral Simian crease, overriding of toes, and syndactyly was seen in three cases each. Polydactyly were found in two children.

An etiology was found in 64 (56.6%) children with GDD. Perinatal asphyxia with hypoxic-ischemic encephalopathy (HIE) was identified as the cause in 15 (13.4%) children. Structural brain malformations were found in 17 children, encephalomalacia in three and periventricular leucomalacia (PVL) in two. Cytoplasmic virus infection and metabolic disorders were found in two children each. The metabolic cause was identified in 44.8% (22/49) GDD children. Among 113 children with GDD, 63 children were identified with CP. Spastic quadriparesis (37) and double hemiplegia (12) were the commonest types. Hypotonic CP was found in six, spastic diplegia in five and extrapyramidal CP in three children.

Among 113 children with GDD, hearing impairment was identified in 35 (30.9%) children (Table 3). Of these 35 children with hearing impairment, 20 (57.1%) children were males and born preterm were 5 (14.2%). History of parental consanguinity was found in 10 (28.5%) children. Microcephaly and dysmorphism were noted in 19 (54.2%) and 11 (31.4%) children respectively.

Among 35 GDD children with hearing impairment, 22 (62.8%) had sensorineural hearing loss, four (11.5%) had conductive hearing loss and nine (25.7%) had combined hearing loss. Among four children with isolated conductive hearing loss, one child had a history of otitis media. Among 31 children with SNHL, 25 (80.6%) had bilateral hearing loss and six (19.4%) had unilateral hearing loss.

Among 31 children with SNHL, hearing loss was severe in 14 (45%) children, moderate in five (16.2%), moderately severe in three (9.6%), and profound in one (3.4%) (Table 4). Nine children had both conductive and sensorineural hearing loss.
Among 35 GDD children with hearing impairment, risk factors other than CP were identified in 10 children (Table 5). Among 63 children with cerebral palsy, 20 (31.7%) had hearing impairment, nine were associated with spastic quadriplegia. The mean developmental quotient in the language domain in children with hearing loss was 38% (SD ± 12.8), the range was 20 - 70%. A hearing aid trial was given to eight children with SNHL during the study period.

DISCUSSION

The present study has identified hearing impairment in 30.9% of children with GDD. Among children with hearing impairment, hearing loss was bilateral in 80.6% and severe in 45%. Because speech and language delay is often associated with GDD and could be the result of a hearing loss, hearing evaluation is mostly undertaken at the first contact.5-12 Children with GDD are at higher risk for hearing loss.7 Early detection of hearing loss at critical developmental stages can prevent or reduce subsequent adverse consequences. In the present study, half of the children (50.5%) were in the age group of six months to 12 months. The mean age at presentation was 13.5 months. The male to female ratio was 1.5:1. In a prospective study by Meena et al. involving 200 children < two years old, 36.8% of GDD children were in the age group of six to 12 months.13 In a study by Tikaria et al. involving 100 children aged < five years, the mean age was 23.6 months.2

We noticed parental consanguinity in 19.5% study population, which is higher compared to 3% in another study.2 Microcephaly was found in 61.1% of GDD children in the present study which is much higher compared to another study which reported microcephaly in 34%.2 We found syndromic features in 2.6% of children which is much lower than those (14%) mentioned by Tikaria et al.2 Dysmorphic features were found in 40 (35.4%) children in our study. Much higher rates (70%) of dysmorphic features were found by Tikaria et al.2

We could find etiology in 56.6% of children with GDD. Tikaria A et al. mentioned etiologic yield in 73% and Srour et al. in 38% of the children.2,14 HIE rates of 13.4% in our study is similar to those found by Tikaria et al. (15%) and Srour et al. (22.4%).2,14 Metabolic cause were identified in 44.8% (22/49) of GDD children in the present study. In other studies, the reported metabolic rates ranges from 2% to 4%.2,13,14 Among 113 children with GDD, 63 children were identified with CP. The higher rate of spastic CP cases in our study is similar to that reported by Singhi et al.15

The hearing impairment in GDD children varies widely.16-25 Hearing impairment was identified in 30.9% of children with GDD in the present study. Rupa et al. reported that 91% of their study children with GDD had hearing loss.16 In contrast, Tikaria et al. reported hearing loss in 20% of study children, and Kwok et al reported hearing loss in 18% among 260 children with severe global developmental delay.2,7 Among 35 GDD children with hearing impairment, 22 (62.8%) had sensorineural hearing loss. Tikaria et al. found SNHL in 20% of children with GDD.2 Angulo et al. found SNHL in 18 (60%) of 30 cerebral palsy children.3 Among the 31 children with SNHL in the present study, 80.6% had bilateral hearing loss and hearing loss was severe in 45% of children. In a retrospective study by Angulo et al. bilateral and unilateral SNHL were reported in 66.6% and 33.3% of children respectively.3

Among 35 GDD children with hearing impairment, risk factors included mainly CP. Among 63 children with cerebral palsy, 20 (31.7%) had hearing impairment, nine were associated with spastic quadriplegia. Angulo et al. reported 60% of children with CP having SNHL. Hearing impairment was noted in 14% of children with CP in another Indian study.15 Other risk factors for hearing loss identified in the present study such as prematurity, kernicterus, CMV infection have been recognized earlier.19,24

We noted that the mean developmental quotient in the language domain in children with hearing loss was 38% (SD ± 12.8). Hearing loss in infants and children may lead to lifelong deficits in speech and language acquisition. Hence, early identification and intervention for children with hearing impairment are suggested for normal children by
Application of such strategies to GDD children is more beneficial. The American Academy of Paediatrics, the American Academy of Neurology and the British Columbia-based Treatable Intellectual Disability Endeavor (TIDE) protocols have proposed multitiered investigations of GDD. These investigations likely guide paediatricians to know the etiology and therapy optimisation. The recommendations also include formal hearing tests in these children. It is also stated that formal hearing testing is critical for all children with suspected GDD. The present study findings agree with these statements. Although our study is a relatively small and single centric study, we presume that our research would help shed more light in this field for the other researchers in the future.

CONCLUSIONS

Hearing impairment is associated with about 30.9% of children with GDD in young children. Isolated sensorineural hearing loss is the predominant type. Among children with SNHL, hearing loss is likely to be bilateral in the majority (80.6%) and severe in about 45%. CP and sequelae of HIE and kernicterus are the predominant risk factors associated with SNHL.

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