

Original Article**Severity of Auditory Impairment in Children with Cerebral Palsy in a Tertiary Health Care Centre**

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Abstract

Background: Cerebral palsy is the most common chronic motor disorder in children and frequently associated with sensorineural pathology. Deafness is very important public health concern. Because it determines long-term life span disability in individuals, decrease their quality of life.

Objectives: To observe the rate of occurrence of hearing impairment in children with cerebral palsy (CP). And to assess the severity of hearing impairment children with CP and correlate the auditory impairment with different types of CP.

Materials and method: In these study 50 children with Cerebral Palsy, on arrival of the CP cases clinical assessment was done. After selecting the CP children who were 1 to 15 years age. A structured questionnaire will be filled up which includes details history of patient and important variables related to hearing status like family history of deafness, consanguinity, antenatal history, preterm LBW, perinatal asphyxia, kernicterus, neonatal septicemia, neonatal seizure and meningitis were included in my study. After taking of details history, physical examination and neurological examination like as hearing, vision, motor, cognitive, speech delay were assessed. Through clinical examination patients were referred to audiologist to do some tests like Tympanometry 'Transient evoke otoacoustic emission (TEOAE), auditory brainstem evoke response (ABR). Tympanometry or immittance Audiometry is used to check the movement of the tympanic membrane. Tones are "bounced" of the tympanic membrane to determine the amount of sound that is reflected back through the ear canal. Middle air pressure, compliance and stepidial reflex threshold (SRT) could be measured by tympanometry. TEOAE is an acoustic reflex looks at the middle ear, inner ear, auditory nerve, brainstem nuclei and facial nerve. An abnormal acoustic reflex can be indicative of a number of pathologies. Transient Evoked Otoacoustic Emission (TEOAE) is reproducible, diagnostically accurate, easy to perform and minimally invasive.

It is important to remember that neither Tympanometry nor Acoustic Reflex tests assess hearing. A person with a normal tympanogram can still have a hearing loss. Hearing assessment would be done by TEOAE. If result was pass, no further follow up would be required in those cases. But those were referred then ABR would be suggested. Severity of hearing impairment could be assessed by ABR. However ABR is costly, time-consuming and requires trained personnel. Since it requires electrode preparation, placement and removal, it is not recommended to be the first choice for hearing assessment.

Results: *In this study, among the 50 children with CP, Tympanometry result negative middle ear pressure were 32% in spastic quadriplegia (SQ), 24% in spastic hemiplegia (SH), 16% in spastic diplegia (SD) and 24% in mixed type. Eustachian tube dysfunction was 62.5% in spastic hemiplegia. Secretory otitis media were 36.4% in spastic quadriplegia and SH. In TOAE result 52% were pass and 48% were refer, results of those were pass, no further follow up hearing assessment would be required. Results of those were refer ABR were suggested. On the basis of result of TOAE, those were referring; initially they were encountered as hearing impairment in this study. Among them, in SQ were 18%, SH were 12% and mixed. Further follow up result of auditory brain stem response revealed mild, moderate and severe sensorineural hearing loss were 8% in spastic quadriplegic type and the spastic diplegia and athetoid group were 2% only mild. Moderate sensori-neural hearing was 2% in spastic hemiplegia group.*

Conclusion: *Hearing loss was frequent in children with Cerebral Palsy. Spastic Quadriplegic cerebral palsy patients had the maximum incidence of hearing impairment. Though the cases of athetoid CP was less but all of them had hearing impairment. Perinatal asphyxia and prematurity were the main risk factor of hearing impairment.*

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Introduction

Cerebral palsy is the 'most prevalent childhood motor disability' today with approximately 8000 infants with cerebral palsy born in the United States annually^{1,2}. The international working group on definition and classification of cerebral palsy, defined cerebral palsy as a group of permanent disorder of development of movement and posture causing activity limitation that are attributed to non progressive disturbances that occurred in the developing fetal or infant brain³.

Most common risk factors are low APGAR score (26.6%) then seizures (13.3%), prematurity (10%) and lastly low birth weight. Intrauterine infections like

cytomegalovirus and rubella, difficult labour kernicterus are least common risk factors⁴.

The prevalence of CP is increased among the low birth weight infants particularly those weighting less than 1000 gm at birth, primarily because of intracerebral hemorrhage and periventricular leukomalacia⁵.

5-10% of children developed cerebral palsy is due to cytomegalovirus⁶. These children suffer from involuntary movement and or disturbance in gait and motility. Children with cerebral palsy may also experience a range of additional problems that may require treatment including impairment of vision, hearing or speech with consequent learning difficulties,

mental retardation, seizure, difficulties in bladder and bowel control and feeding problem⁴.

Hearing loss account for 25% of cerebral palsy population in Western Europe. These abnormalities are thought to be associated with prenatal, perenatal and postnatal problems often multifactorial in nature⁷.

In persons with cerebral palsy especially in the affected group, incidence of hearing impairment has been reported at 10- 41%. Out of 1649 cases of cerebral palsy patient 12 had severe hearing disability in England and Scotland. Hearing loss was averaged across frequencies 0.5 - 4 KHz and the disability classified as “mild” if hearing loss was 21- 45 dB, “moderate” for 45 - 70 dB loss, “severe” with a hearing loss >70 dB. Assessment of accurate hearing levels was often impossible in children with concomitant intellectual impairment⁸. Eighty percent of preterm infants developed hyperbilirubinaemia during first week of life. Additionally, this preterm population is at a higher risk to develop secondary disorder as a result of the permeability of the blood brain barrier. Severe kernicterus usually lead to death of infants with in first few weeks of life. When the infant survives the kernicterus, athetoid type of CP is often seen with hearing loss and cognitive impairment⁹. Intrauterine infection, ototoxic drugs, bacterial meningitis, hypoxia, chromosomal abnormalities can cause both CP and hearing loss¹⁰. Sensorineural hearing loss (SNHL) is found in a small proportion of very preterm survivors¹¹.

There are many ways to detect hearing impairment in children. To formulate a protocol for infant hearing screening in developing countries enabling it to be later incorporated into their national deafness screening programs. The screening tool should be sensitive in detecting hearing loss in infants with high specificity¹². Tympanometry or immittance Audiometry is used to

check the movement of the tympanic membrane. Tones are "bounced" of the tympanic membrane to determine the amount of sound that is reflected back through the ear canal. Middle air pressure, compliance and stepidial reflex threshold (SRT) could be measured by tympanometry. Transient Evoked Otoacoustic Emission (TEOAE) is reproducible, diagnostically accurate, easy to perform and minimally invasive¹³. TEOAE represents a quick, inexpensive, frequency specific, valid method for testing cochlear function but it can be misleading in case of wax packed ear canal, middle ear effusion, external noise as these conditions often confound the real audiological output. Moreover TEOAE cannot identify auditory neuropathy¹⁴.

Brainstem Evoked Response Audiometry (BERA), however, is not sensitive to such conditions mentioned above and can identify auditory neuropathy. However BERA is costly, time-consuming and requires trained personnel. Since it requires electrode preparation, placement and removal, it is not recommended to be the first choice for screening of hearing-impaired child. BERA is indicated when two consecutive TEOAE assessments are inconclusive¹⁵.

Materials & Methods

This cross-sectional study was conducted in the Department of Pediatrics neurology unit and Autolaryngiology – Head and Neck Surgery Department of BSMMU. Duration of the study was from March 2011 to September 2011. Diagnosed CP patients who fulfils the inclusion and Exclusion criteria was selected purposively as study population. After taking informed written consent, proper history and clinical examinations was done and severity of hearing loss was assessed by different tools of hearing in the auditory department of Autolaryngiology – Head and Neck Surgery.

Result

Table 1: Demographic characteristics of study population (n=50)

Characteristics	Frequency	Percentage (%)
Sex		
Male	32	64.0
Female	18	36.0
Age group		
1-3 years	22	44
3-6 years	16	32
6-15 years	12	24
Socio -economic status		
Poor	26	52.0
Middle class	22	44.0
Higher class	02	04.0
Residence		
Rural	27	54.0
Urban	23	46.0

Table-1 shows male was 64% and female was 36% of study population. 44% children were 1-3 years age group, 32% children were 3-6 years age group and 24% children were 6-15 year's age group. Mean age in year 5.57 ± 3.89 , age range 1-15 years. Regarding socioeconomic status, poor was 52%, middle class was 44% and higher class was 04%. 54% children come from rural area, and 46% children come from urban area.

Table 2: Type of CP in the study population (n=50)

Type of CP	Frequency	Percentage (%)
Spastic Quadriplegia (SQ)	21	42.0
Spastic Hemiplegia (SH)	11	22.0
Spastic Diplegia (SD)	08	16.0
Athetoid	02	04.0
Mixed	08	16.0
Total	50	100.0

Table- 2 shows out of 50 patients 42%, were spastic quadriplegia, 22% were spastic hemiplegia, 16% were spastic diplegia, 04% were athetoid and 16% were mixed CP.

Table- 3: Tympanometry report of various type of CP (n=50)

Tympanometry	Type of CP					Total n (%)
	SQ	SH	SD	Mixed	Athetoid	
	n (%)	n (%)	n (%)	n (%)	n (%)	
Negative middle ear pressure	8(32)	6(24)	4(16)	6(24)	1(4)	25 (50)
Eustachian tube dysfunction	2(25)	5(62.5)	1(12.5)	0(0)	0(0)	08 (16)
SRT (Absent)	4(36.4)	4(36.4)	0(0)	2(18.2)	1(9.1)	11 (22)

Coefficient value 0.40, (p=0.12)

Table 3 Tympanometry result shows negative middle ear pressure was 32%, Eustachian tube dysfunction was 25% in SQ .24% was negative middle ear pressure, 62.5% was Eustachian tube dysfunction in SH, SRT was absent in 36.4% in SQ and SH. (P=0.12) that was not statistically significant.

Table 4: Etiology of CP among the study population (n=50)

Etiology	Frequency	Percentage (%)
Perinatal asphyxia	26	52
Prematurity	17	34
Meningitis	12	24
Neonatal septicemia	11	22
Brain malformation	08	16
Kernicterus	02	04

Table 4 Shows etiology in the study population perinatal asphyxia were 52%, prematurity were 34%, meningitis were 24%, neonatal septicemia were 22%, brain malformation were 16, kernicterus were 4%.

Table 5: Hearing status by TEOAE in various type of CP (n=50)

Type of CP	Number of Children	Refer N (%)	P value
SQ	21	09 (18)	0.25
SH	11	06 (12)	0.01
SD	08	03(06)	0.12
Athetoid	02	02 (04)	1.0
Mixed	08	04(08)	0.03
Total	50	24(48)	

Chi -Square test

Table- 5 TEOAE result shows hearing impairment of various type of CP. Among them, in SQ were 18%, SH were 12%, (p = 0.01) that was statistically significant. And mixed were 8%, (p= 0.03) that was statistically significant

Table 6: Level of hearing impairment by Auditory Brainstem Response (ABR) in various type of CP (n=50)

Type of CP	Normal	Mild hearing loss	Moderate hearing loss	Severe to profound hearing loss	Total sensorineural hearing loss
Spastic quadriplegia	05(10%)	02(4%)	01(2%)	01(2%)	04 (8%)
Spastic hemiplegia	05(10%)	00	01(2%)	00	01(2%)
Spastic diplegia	02(10%)	01(2%)	00	00	01(2%)
Athetoid	01(4%)	01(2%)	00	00	01(2%)
Mixed	04(2%)	00	00	00	00

Coefficient value 0.75, p value – 0.15

Table 6 shows result of auditory brain stem response revealed mild, moderate and severe to profound sensorineural hearing loss were 8% in spastic quadriplegic type, moderate sensorineural hearing loss was 2% in spastic hemiplegic group and mild hearing loss was 2% in spastic diplegic and athetoid group of CP. (P=0.15) that was not statistically significant.

Discussion

This descriptive study was conducted in the Department of Pediatrics (Pediatric Neurology unit) BSMMU. This study was carried out to observe the Frequency and Severity of Auditory Impairment in children with Cerebral Palsy.

It has been noted that roughly 20 percent or one fifth of children with cerebral palsy suffer from hearing loss. Typically the hearing loss is sensorineural. Beyond that, not much research has been done to date on hearing loss and cerebral palsy. It is important, however, that children with cerebral palsy also have their hearing assessed by an audiologist, especially if they also have a visual impairment.

A large number of those with CP will have additional impairments, such as visual, auditory, seizures, cognitive and behavioral disabilities⁶.

Of those with hearing loss 30.2% will have an additional disability¹¹. Cerebral palsy can have associated disabilities, hearing loss, visual impairments, epilepsy, speech and language disorders and cognitive impairments⁶.

It is a hidden disability for which early detection is usually missed. But early detection and intervention can prevent severe psychosocial, educational and linguistic repercussions.

Out of 50 children, thirty two (64%) were male and eighteen (36%) were female. 44% children were 1-3 years age group, 32% children were 3-6 years age group and 24% children were 6-15 years age group. Mean age 5.57 years, SD±3.89 age range 1-15 years.

In the study of Liptak GS, et al¹⁶ mean age was 9.6 years SD ±4.6 that is dissimilar in this study, 59% were male, 41% were female that is approximately similar with this study.

In this study, normal delivery was 78 % and caesarean section was 22% in children with CP. In study of

Mezaal M A, et al.¹⁷ 82% were normal delivery and 18% were caesarean section. That study is similar with this study.

In the study of hearing impairment was associated with preterm low birth weight 34%, perinatal asphyxia 52%, severe Jaundice 04%, neonatal septicemia was 22% and neonatal seizure were 24%.

In the study of Shilpi Arora and LK Kochher¹⁸ risk factors for hearing loss was low birth weight 41.42% which near to similar, hyperbilirubinemia 25.71%, septicemia 4.28% and asphyxia 7.14% that is dissimilar in my study.

In the study of Behairy E H. et al.⁴ represents the associated problems in their cases which are from the most common to the least common; seizures (63.3%) auditory (53.3%), speech (46.7%), visual (46.7%) and lastly swallowing problems in (16.6%) of the cases.

Associated problems in this study motor delay were forty two (84%), speech delay were thirty nine (78%), seizure were thirty five (70%), hearing impairment were twenty nine (58%), cognitive delay were twenty nine (58%), visual impairment were twenty (40%), swallowing difficulty were nineteen (38%) and dental problem were fourteen (28%). This study is approximately similar Behairy E H.⁴ study.

In another study Mezaal M A et al,¹⁷ motor disabilities was 60%, most common followed by epilepsy 42%, hearing loss 63%, cognitive delay 40%, speech delay 30% that study approximately similar to in this study. In Mezaal M A et al,¹⁷ study visual impairment 2% that is distinct in this study.

In this study out of 50 patients, 42% were spastic quadriplegia, 22% were spastic hemiplegia, 16% were spastic diplegia, 18% were mixed CP.

In the study of Behairy E H. et al.⁴ demonstrates 76.6% were spastic quadriplegia which nearly similar,

6.67% were spastic hemiplegia and 3.33% were spastic diplegia which is not similar in this study.

In study of Mezaal M A et al,¹⁷ 32% were spastic hemiplegia, 18% were spastic quadriplegia which was dissimilar but 12% were spastic diplegia, 16 % were mixed CP that was almost similar with this study.

In this study, Tympanometry result shows negative middle ear pressure (secretory otitis media) was 32%, Eustachian tube dysfunction was 25% (combined were 57%). 24% was negative middle ear pressure, 62.5% was Eustachian tube dysfunction in SH, SRT was absent in 36.4% in SQ and SH.

In the study of Behairy E. H. et al,⁴ found that, abnormal tympanometry in the form of secretory otitis media, eustachian tube dysfunction, was present in 52% of spastic quadriplegic patients while other types of cerebral palsy had normal tympanometry. This is similar to this study.

In this study, TEOAE report pass was 52% and refer was 48% in children with CP. Theodore et. al showed in their study pass 73,4% and refer 26.2% which is not consistent with in this study.

In this study auditory brain stem response revealed mild, moderate and severe sensorineural hearing loss were 8% in spastic quadriplegic type and the spastic diplegia and athetoid group were 2% only mild. Moderate sensorineural hearing loss were 2% in spastic hemiplegia group.

In the study of Behairy E. H. et al,⁴ found that result of auditory brain stem response revealed mild sensorineural hearing loss in moderate and severe spastic quadriplegic type. Athetoid group only mild. Moderate sensorineural hearing in spastic hemiplegia group. That is similar to in this study.

Conclusion

Hearing loss was frequent in children with Cerebral Palsy. Management of this problem is difficult because

of the motor disorder and the associated pathology. Early audiological assessment is very important to improve the language outcome in these children. Spastic Quadriplegic cerebral palsy patients had the maximum incidence of hearing impairment. Though the cases of athetoid CP were less but all of them had hearing impairment. Perinatal asphyxia and prematurity were the main risk factor of hearing impairment. So early diagnosis of CP would be helpful for prevention of disability of some problems.

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