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Original Research

Auditory neuropathy with hearing loss in children

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ABSTRACT:

Background: Auditory neuropathy/dyssynchrony (AN/AD) is a hearing disorder characterized by an absent or atypical auditory brainstem response. The present study was conducted to determine cases of Auditory neuropathy/dyssynchrony with hearing loss. **Materials & Methods:** 140 patients diagnosed with hearing loss age ranged 1- 14 years were subjected to hearing screening tests. The prevalence of AN/AD was assessed. **Results:** Out of 140 patients, boys were 80 and girls were 60. Out of 140 patients, 7 (5%) had AN/ AD. Acoustic reflex was present in 7, high Bilirubin was present in 6, blood exchange after birth was seen in 5 and neonatal intensive care was required in 6. The difference was significant (P<0.05). **Conclusion:** Auditory neuropathy in children was associated with hyperbilirubinemia and high acoustic reflex.

Key words: Auditory neuropathy, Children, hearing

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INTRODUCTION

Auditory neuropathy/dyssynchrony (AN/AD) is a hearing disorder characterized by an absent or atypical auditory brainstem response (ABR), with preservation of the cochlear microphonics (CM) and/or otoacoustic emissions (OAEs).[1] The prevalence accounts vary from roughly 1% to 10% in schools for the deaf and between 10% in newborns and 40% in hearing-impaired neonatal intensive care unit (NICU) graduates.[2] The authors found that these patients were most likely alike to those previously reported cases with a paradoxical absence of ABRs and only a slight impairment of pure tone thresholds but in whom CMs or OAEs had not been recorded.[3] AN/AD is characterized by a unique pattern of hearing loss and distorted ABR with preservation of outer hair cell function.[4] AN/AD comprises a spectrum of pathology affecting the auditory pathways anywhere from the inner hair cells to the brainstem. Thus it is difficult to define the disorder as cochlear or retrocochlear. Increased clinical suspicion supported by appropriate diagnostic tests is needed to establish an accurate diagnosis.[5]

It has been established that primary detection of these patients is very helpful in rehabilitation and instruction. AN/AD was identified in the pediatric population by initiating neonatal hearing screening.[6] The present study was conducted to assess cases of Auditory neuropathy/dyssynchrony with hearing loss in children

MATERIALS & METHODS

The present study comprised of 140 patients diagnosed with hearing loss age ranged 1- 14 years of both genders. All patients were informed regarding the study and parental written consent was obtained.

Data such as name, age etc. was recorded. All patients were subjected to hearing screening tests such as tympanometry, TEOAE, DPOAE, AABR by an expert ENT surgeon. The prevalence of AN/AD was assessed. Results thus obtained were subjected to statistical analysis. P value less than 0.05 was considered significant.

RESULTS

Table I Distribution of patients

Total- 140		
Gender	**Boys**	**Girls**
Number	80	60

Table I shows that out of 140 patients, boys were 80 and girls were 60.

Table II Prevalence of AN/AD

Total	Prevalence	Percentage
140	7	5%

Table II shows that out of 140 patients, 7 (5%) had AN/ AD.

Table III Assessment of parameters

Parameters	Variables	Number	P value
Acoustic reflex	Yes	7	0.01
	No	0	
High Bilirubin	Yes	6	0.03
	No	1	
Blood exchange after birth	Yes	5	0.05
	No	2	
Neonatal intensive care unit	Yes	6	0.03
	No	1	

Table III shows that acoustic reflex was present in 7, high bilirubin was present in 6, blood exchange after birth was seen in 5 and neonatal intensive care was required in 6. The difference was significant (P<0.05).

DISCUSSION

The function of the external hair cells of the cochlea is normal, while the neural conduction at the level of the vestibulo-cochlear is damaged.[7] That's why children and infants with this neurological disorder are not detected using OAE.[3] Because, it only examines hearing to the earliest level of the ear or the hair cells of the cochlea and this lack of identification causes a wide range of the problems, including language, communication and educational difficulties.[8]

The clinical findings for auditory neuropathy are associated with several diagnoses including hyperbilirubinemia, neurodegenerative diseases,[9] Charcot-Marie-Tooth syndrome, and other sensorimotor neuropathologies, mitochondrial disorders and ischemic-hypoxic neuropathy resulting from asphyxia. Also, experimental animal models for auditory neuropathy have been proposed using the carboplatin ototoxicity and ischemic-hypoxic neuropathy methodologies.[10] The present study was conducted to determine cases of Auditory neuropathy/dyssynchrony.

We found that out of 140 patients, boys were 80 and girls were 60. Berlin et al[11] included 42 patients, 21 (50%) were in the age group of 11–20 years followed by 13 patients who were between 0 and 10 years (30.95%). The remaining 8 were aged above 20 years (19.04%). The youngest patient was 10 months old and the oldest was aged 38 years with a mean age of 10.35 ± 2.10 years. There were 29 (69.04%) females and 13 (30.95%) males. 3/42 (7.14%) patients gave a history of exposure to ototoxic drugs such as streptomycin, gentamicin, and kanamycin, but never had a history of loss of hearing before that. History of premature birth was noted in 10 (23.80%) patients and the remaining patients did not show premature birth history. Among the 42 patients of this study group, 23 (54.76%) had low birth weight, of which 2/42 (4.76%) were <1.5 kg. Authors suggested that The major risk factor for ANSD was low birth weight with prematurity, NICU admissions, and viral infections having significant contributions.

We observed that out of 140 patients, 7 (5%) had AN/ AD. Mittal et al[12] in their study found that 40% of patients had hyper bilirubin level, 13.3% had oxygen deficiency and 46.7% of patients had no specific disease history. In a study, which was on the patients in NICU, were observed that in comparison with normal children, there is the predisposition factors for AN/ AD children, such as IRDS, meningitis and the use of vancomycin.

We found that acoustic reflex was present in 7, high Bilirubin was present in 6, blood exchange after birth was seen in 5 and neonatal intensive care was required in 6. Uhler et al[13] enrolled 105 hearing impairment children. All them were under hearing screening tests. 4 cases (8 ears) with AN/ AD were diagnosed, which had an average age 37 months. The prevalence of AN/AD was 3.8 % among hearing impaired children. The findings of this study showed that there are the relationships between AN/ AD and fluctuating hearing loss, acoustic reflex, high bilirubin, blood exchange after birth, neonatal intensive (NICU) care unit. The simultaneous use of both ABR and OAE tests in the birth screening provide much more useful information than when each of these tests is used alone.

The limitation of the study is small sample size.

CONCLUSION

Authors found that auditory neuropathy in children were associated with hyperbilirubinemia and high acoustic reflex.

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