

ORIGINAL ARTICLE

Evaluation of cases of Auditory neuropathy/dys-synchrony with hearing loss- A clinical study

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ABSTRACT:

Background: Auditory neuropathy/dys-synchrony (AN/AD) is a hearing disorder characterized by an absent or atypical auditory brainstem response. The present study was conducted to evaluate cases of Auditory neuropathy/dys-synchrony with hearing loss. **Materials & Methods:** The present study was conducted on 218 patients diagnosed with hearing loss age ranged 1- 14 years of both genders. All patients were subjected to hearing screening tests (tympanometry, TEOAE, DPOAE, AABR). The prevalence of AN/AD was assessed. **Results:** Out of 218 patients, 8 (3.67%) had AN/ AD. Acoustic reflex was present in 8, high Bilirubin was present in 7, blood exchange after birth was seen in 6 and neonatal intensive care was required in 6. The difference was significant ($P < 0.05$). **Conclusion:** Authors found that cochlear-sensitive auditory test (OAE) and the auditory brain stem response (ABR) is necessary for the early diagnosis of AN/AD.

Key words: Auditory neuropathy, cochlear-sensitive auditory test, hearing

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INTRODUCTION

Auditory neuropathy/dys-synchrony (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 function of the external hair cells of the cochlea is normal, while the neural conduction at the level of the vestibulo-cochlear is damaged.²

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.⁴ 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. Starr et al first described this rare condition. 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.⁵

It has been established that primary detection of these patients is very helpful in rehabilitation and instruction. The term of AN/AD was first proposed in the 1980s, following the observation of the patients with normal pure tone thresholds, which had difficulty to detect the sounds, especially in a noisy environment. AN/AD was identified in the pediatric population by initiating neonatal hearing screening.⁶ The present study was conducted to determine cases of Auditory neuropathy/dys-synchrony with hearing loss.

MATERIALS & METHODS

The present study was conducted in the department of ENT. It comprised of 218 patients diagnosed with hearing loss age ranged 1- 14 years of both genders. Ethical approval was obtained from institute prior to the study. All patients were informed regarding the study and written consent was obtained.

Data such as name, age etc. was recorded. All patients were subjected to hearing screening tests (tympanometry, TEOAE, DPOAE, AABR). 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- 218		
Gender	Male	Female
Number	120	98

Table I shows that out of 218 patients, males were 120 and females were 98.

Table II Prevalence of AN/AD

Total	Prevalence	Percentage
218	8	3.67

Table II, graph I shows that out of 218 patients, 8 (3.67%) had AN/ AD.

Graph I Prevalence of AN/AD

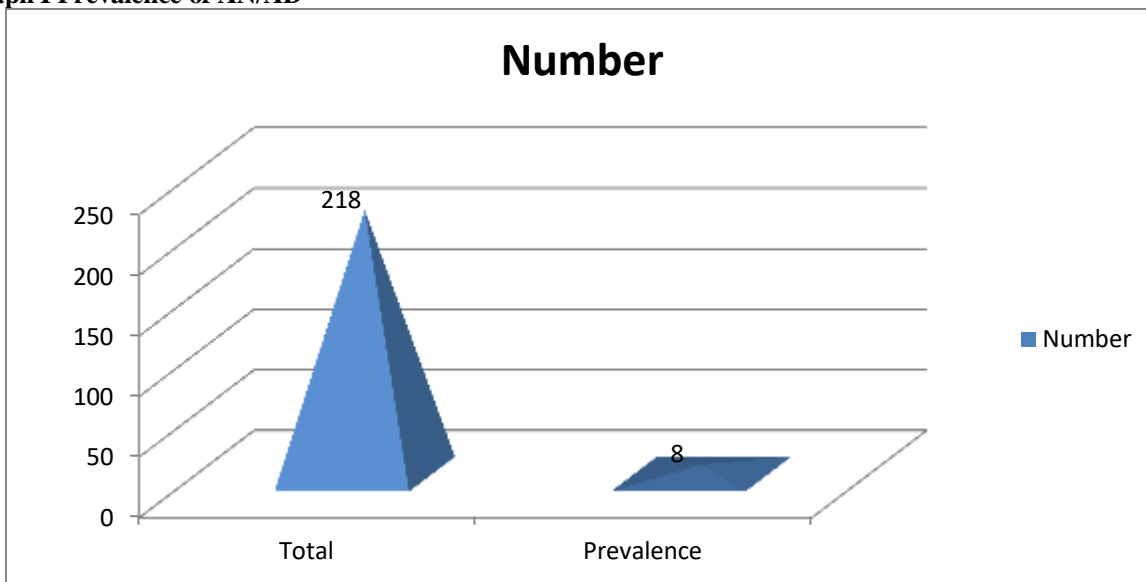


Table III Assessment of parameters

Variables	Number	P value
Acoustic reflex		
Yes	8	0.01
No	0	
High Bilirubin		
Yes	7	0.02
No	1	
Blood exchange after birth		
Yes	6	0.05
No	2	
Neonatal intensive care unit		
Yes	7	0.02
No	1	

Table III shows that acoustic reflex was present in 8, high Bilirubin was present in 7, blood exchange after birth was seen in 6 and neonatal intensive care was required in 6. The difference was significant (P<0.05).

DISCUSSION

AN/AD is characterized by a unique pattern of hearing loss and distorted ABR with preservation of outer hair cell function.⁷ 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.⁸

The clinical findings for auditory neuropathy are associated with several diagnoses including hyperbilirubinemia,⁹ neurodegenerative diseases,¹⁰ Charcot-Marie-Tooth syndrome, and other sensorimotor neuropathologies, mitochondrial disorders and ischemic-hypoxic neuropathy resulting from asphyxia.¹¹ Also, experimental animal models for auditory neuropathy have been proposed using the carboplatin ototoxicity and ischemic-hypoxic neuropathy methodologies.¹² The present study was conducted to determine cases of Auditory neuropathy/dys-synchrony.

In present study, out of 218 patients, males were 120 and females were 98. Uhler et al¹³ conducted a study which consisted of 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. So, 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.

We found that out of 218 patients, 8 (3.67%) had AN/AD. Acoustic reflex was present in 8, high Bilirubin was present in 7, blood exchange after birth was seen in 6 and neonatal intensive care was required in 6. Berlin et al¹⁴ 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.

Mittal et al¹⁵ 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.

CONCLUSION

Authors found that cochlear-sensitive auditory test (OAE) and the auditory brain stem response (ABR) is necessary for the early diagnosis of AN/AD.

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