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Original Article

Audiological Profile in Auditory Neuropathy Spectrum Disorder – A Descriptive Study

Surendran Kizhakkayil Meethal[1], K P Sunil Kumar[2], P Muraleedharan Nampoothiri[2], Slimcy Shilen[3]

[1]Associate Professor, Department of ENT, Government Medical College, Kozhikode, Kerala, India, [2] Professor, Department of ENT, Government Medical College, Kozhikode, Kerala, India, [3]Senior Resident, Department of ENT, Government Medical College, Kozhikode, Kerala, India

Abstract

Background: Auditory neuropathy, auditory dys-synchrony, and auditory neuropathy spectrum disorder (ANSD) are variable terms used to describe an auditory disorder seen in patients ranging in age from infants to adults. The prevalence of ANSD in deaf schoolchildren is 2.46% within the age range of 6–12 years. In children, they are detected by the presence of otoacoustic emissions (OAEs) in the absence of ABRs. In older age group, difficulty hearing in noise, fluctuating hearing, and speech perception performance not predict ed by the level of residual hearing have been reported. The multitude of etiologies for ANSD results in heterogeneous group of patients – making the management strategies even more challenging. The common etiologies put forward are – prematurity, neonatal insult, genetic abnormality, ototoxic drugs, and head injury.

Aim of the Study: This study aims to study the audiological profile in ANSD in a tertiary care hospital and to study the etiology of ANSD cases.

Materials and Methods: A total of 42 patients attending the ENT Outpatient Department of Government Medical College, Kozhikode, with ANSD were included in the study. An ethical committee clearance was obtained before the commencement of the study. An ethical committee cleared consent form was used for the study. All ANSD patients were evaluated with a detailed history including perinatal and development history, ototoxic drug exposure, head trauma, neurodegenerative conditions, and family history. Following clinical evaluation which included general examinations, ENT examination, and central nervous system examination, an audiological evaluation, which included pure tone audiometry, speech audiometry, immittance evaluation, OAE, and auditory brainstem response, was done. Radiological investigation (magnetic resonance imaging brain with inner ear – focusing on any structural anomalies; cochlea, vestibulocochlear nerve, and internal auditory canal) was done. Patients were counseled regarding the rehabilitation options based on their audiological and radiological results and the need for follow-up was explained.

Observation and Results: A total of 42 patients attending the ENT Outpatient Department (OPD) of Government Medical College, Kozhikode, with ANSD were included in the study. Among the 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. 21/42 (50%) patients had birth weight above 1.5 kg. 10/42 patients (23.80%) gave a history of neonatal intensive care unit (NICU) admissions at the time of their birth.

Conclusions: The major risk factor identified in this study for ANSD was low birth weight with prematurity, NICU admissions, and viral infections having significant contributions. On audiological evaluation, hearing loss was of mild-to-moderate range with a low-frequency loss. There was no statistical correlation between pure audiometry values and speech audiometry which was a characteristic observation. OAEs were present in the majority of patients with absent cochlear microphonics (reverse polarity) and acoustic reflexes.

Key words: Auditory dys-synchrony, Auditory neuropathy spectrum disorder, Auditory neuropathy

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INTRODUCTION

Auditory neuropathy/auditory dys-synchrony/auditory neuropathy spectrum disorders (ANSDs) describe a condition, in which patient's otoacoustic emissions (OAEs) are (or were at 1 time) present, and auditory brainstem responses (ABRs) are abnormal or absent.[1] The first

Corresponding Author: Dr. K P Sunil Kumar, Department of ENT, Government Medical College, Kozhikode, Kerala, India.

audiological report of ANSD was probably by Hinchcliffe *et al.*,[2] Starr *et al.*[3] introduced the term "neuropathy" after studying 10 patients with a unique set of auditory problems. In 2001, Berlin *et al.*[4] introduced the term auditory neuropathy (AN)/auditory dys-synchrony to include those cases where no true neuropathy was apparent when the constellation of routine test results did not provide sufficient evidence to differentiate between synaptic dysfunction and "true neuropathy" of the cochlear nerve. Rance[5] studied children with AN and reported that half of them had speech perception abilities like those of children with matched sensorineural hearing loss (SNHL) and had cortical evoked potentials. The other half did poorly in speech reception and lacked cortical responses. In the current terminology, ANSD is a disorder characterized by the disruption of the temporal coding of acoustic signals in the auditory nerve fibers, resulting in the impairment of auditory perceptions that rely on temporal cues.[6] The prevalence accounts vary from roughly 1%[7] to 10% in schools for the deaf and between 10% in newborns and 40% in hearing-impaired neonatal intensive care unit (NICU) graduates.[3] Diagnosis usually needs a high index of suspicion. Although a detailed history might give a clue to some derangement, it is not uncommon to see patients diagnosed by audiological evaluation alone. On the one hand, patient's will report with normal to severely compromised hearing and at the other extreme cases of ANSD presenting as treatment-resistant anxiety disorder can be seen.[8] The late-onset ANSD can be a quite debilitating condition as the clients are perfectly normal till adolescence and suddenly exhibit auditory symptoms. This leads to poor communication among the peer groups and social isolation and decline in academic performance. All these lead to psychological issues such as stress depression and anxiety in persons with late-onset ANSD.[9] There is no definite evidence to pinpoint to site of lesion in ANSD. It could be anywhere beyond the outer hair cells. It will very difficult to categorize the entity into cochlear or retrocochlear.[10] There is no single test for the localization of the site of the lesion. In some cases, the damage might be to the inner hair cells; in other cases, the cause may involve damage to the auditory neurons that transmit sound information from the inner hair cells to the brain. Combinations of these problems might occur in some cases.[11] Rance and Starr pointed out that presynaptic and postsynaptic disorders can cause an ANSD phenotype. The presynaptic disorders include inner hair cell dysfunction and/or loss and deficits in the neurotransmitter release from the inner hair cell-dendrite synapse. The postsynaptic disorders include unmyelinated dendritic nerve terminals dysfunction, axonal neuropathies, auditory ganglion cell disorders,[12] demyelination disorders, auditory nerve hypoplasia, and auditory nerve conduction block.[12] Starr *et al.*[3] suggested an etiological classification of ANSD based on this: Type I postsynaptic AN plus vestibular and peripheral neuropathies and Type II postsynaptic AN plus optic nerve disorders accompanying nuclear and mitochondrial mutations. Type III presynaptic AN plus inner hair cell and neurotransmitter disorder and Type IV auditory neuropathy unspecified where the affected sites are unknown.

Type of Study

This was a cross-sectional, descriptive, and analytical study.

Institute of Study

This study was conducted at Government Medical College and Hospital, Kozhikode.

Period of Study

This study was from January 2017 to June 2018.

MATERIALS AND METHODS

A total of 42 patients attending the ENT OPD of Government Medical College, Kozhikode, with ANSD were included in the study. An ethical committee clearance was obtained before the commencement of the study. An ethical committee cleared consent form was used for the study.

Inclusion Criteria

(1) Patients with a history and clinical examination suggestive of ANSD were included in the study. (2) Patients of all age groups were included in the study. (3) Patients of both the genders were included in the study.

Exclusion Criteria

(1) Patients who failed to complete audiological evaluation were excluded from the study. (2) Patients with a previous history of inflammatory diseases of the ears were excluded from the study. (3) Patients with uncontrolled diabetes and thyroid deficiencies were excluded from the study. All ANSD patients were evaluated with a detailed history including perinatal and development history, ototoxic drug exposure, head trauma, neurodegenerative conditions, and family history. Following clinical evaluation which included general examinations, ENT examination, and central nervous system examination, an audiological evaluation, which included pure tone audiometry (PTA), speech audiometry, immittance evaluation, OAE, and ABR was done. Radiological investigation (magnetic resonance imaging [MRI] brain with inner ear – focusing on any structural anomalies; cochlea, vestibulocochlear nerve, and internal auditory canal) was done. Patients were counseled regarding the rehabilitation options based on their audiological and radiological results and the need for follow-up was explained. All the data were analyzed with standard statistical methods.

Sample Size

The sample size was 42 (based on the previous hospital records and statistical analysis); $n = 4\ pq/d^2$ wherein p=60%, q=100-60=40%, d=15% $n = 4 \times 60 \times 40/152$ and hence $n = 42$.

Working Strategy

(A) Patients were evaluated and diagnosed using the following methods: (1) Proper history and presenting complaints. (2) Intake of ototoxic drugs, associated neurodegenerative disorders or head injury was elicited. (3) Perinatal history and familial causes were noted. (B)(1) Patients were subjected to general examination and ENT examination, central nervous system examination – for any neurodegenerative causes. (2) Audiological evaluation including pure tone audiometry, speech audiometry, immittance evaluation, OAE, and ABR was done. (3) Radiological evaluation of the inner ear with MRI was done. (4) Following the study, the results were analyzed and rehabilitation was done.

OBSERVATION AND RESULTS

A total of 42 patients attending the ENT OPD of Government Medical College, Kozhikode, with ANSD were included in the study. Among the 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 [Table 1]. There were 29 (69.04%) females and 13 (30.95%) males [Table 1].

Among the 42 patients, 41 (97.61%) had bilateral hearing loss and the remaining 1 patient had (2.38%) unilateral hearing loss and it was on the left side. None of the patients had hearing loss in their right ear [Table 2].

The incidence of tinnitus was observed in this study group and found that 30 (71.42%) of them did not complain of any tinnitus. Ten (23.80%) patients complained of tinnitus in their both ears and 2 patients (4.76%) had tinnitus in their right ear [Table 3].

Among the 42 patients, 36 (85.71%) had no complaints of vertigo and 6 (14.28%) had vertigo. All the 42 patients (100%) presented with poor speech discrimination (excluding children <5 years). Clarity of the speech was good in 25 (59.52%) of the patients. Clarity of the speech was poor in 17 (40.47%) of the patients [Table 4].

Examining the various etiologies that could possibly the causing ASND in this study, it was observed that 14 (33.33%) patients had a history of systemic viral infections before the onset of the disease. Of 14 patients with viral infections, 7 patients had suffered from viral parotitis (16.66%). The remaining 28/42 patients showed no history or ailment of viral infections before ASND disease [Table 5].

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

Table 1: The distribution of the patients according to their age groups and gender (*n*=42)

Age groups in years	Number (%)	Males (%)	Females (%)
0–10	13 (30.95)	3 (7.14)	10 (23.8)
11–20	21 (50)	7 (16.66)	14 (33.33)
21–30	5 (11.9)	2 (7.14)	3 (11.9)
Above 30	3 (7.14)	1 (2.38)	2 (4.76)

Table 2: The involvement of side of hearing loss in the study group (*n*=42)

Side of hearing loss	Number (%)
Unilateral	41 (97.61)
Bilateral	1 (2.38)
Right ear	42 (100)
Left ear	0 (0)

Table 3: The incidence of tinnitus in the study (*n*=42)

Tinnitus	Number (%)
Absent	30 (71.42)
Right ear	10 (23.8)
Left ear	2 (4.76)

Table 4: The incidence of vertigo, speech discrimination, and clarity of speech in the study (*n*=42)

Symptom	Present (%)	Absent (%)
Vertigo	36 (85.71)	6 (14.28)
Poor discrimination of speech	42 (100)	0 (0)
Clarity of speech	25 (59.52)	17 (40.47)

Table 5: The incidence of viral etiology in the study population (*n*=42)

Etiological factors	Number	Percentage
Viral infections-14		
Other than viral parotitis	7	16.66
Viral parotitis	7	16.66
Total		33.33
No viral infections	28	66.66

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. 21/42 (50%) patients had birth weight above 1.5 kg. 10/42 patients (23.80%) gave a history of NICU admissions at the time of their birth. 5/42 (11.9%) of the patients in this study had a history of head trauma. Consanguinity was observed in 5/42 patients gave a history of having born out of (11.9%) their parent's consanguineous marriage. Peripheral neuropathy was observed in 4/42 (9.50%) of the patients had peripheral neuropathy and 90.5% had no evidence of peripheral neuropathy. Of the 42 patients studied, only 1 had (2.38%) neurodegenerative disease – hereditary spastic paraplegia. To summarise the etiological factors playing a role in causing ANSD in patients of this study, it was observed that. Of the 42 patients studied, the major association was found with low birth weight (54.8%), followed by viral infection (33.3%), NICU admission (23.8%), and prematurity (23.8%). Some of them had more than one causative factor. The least common was neurodegenerative disease (2.4%). About 30.95% of them had no identifiable cause [Table 6].

On clinical examination of the ear, it was observed that 9 (21.42%) of the study participants had retracted (Grade I) tympanic membrane in both ears, whereas 78.6% had intact tympanic membrane in both ears. In the study population of 42 patients, 22 had moderate hearing loss (52.38%). Hearing loss was mild in 6 (14.28%) patients in their right ears [Table 7].

In the study population of 42 patients, 14 had moderate hearing loss (33.33%). Hearing loss was mild in 14 (33.33%) patients in their left ears [Table 8].

Looking at the pattern of pure tone audiometry and loss of hearing across the frequencies, it was observed that 90.4% had low-frequency type of loss and 4.8% had flat and the remaining 4.8% had high-frequency loss. High-frequency loss was noted in patient with hereditary spastic paraplegia. Analysis of the speech audiometry in the study subjects showed that all (100%) the ANSD ears had poor speech discrimination score <25%. Speech reception threshold was more than 45% among the 38/42 (90.47%) of the patients. All (100%) of the patients with ANSD had poor correlation between PTA and speech audiometry, $P = 0.153$ (where P was significant at <0.05). Similarly, there was poor correlation between pure tone average and speech discrimination score with $P = 0.701$ (where P was significant at <0.05). Tympanometry studies of the patients with ANSD were carried out and it was analyzed that 39/42 (92.85%) had normal tympanograms (Type A) and the remaining 7/42 (7.14%) had C type of tympanogram. All the patients (100%) had absent acoustic reflexes on impedance audiometry. The study of OAEs in the present study showed that 36/42 (85.71%) had OAEs present in their right ears and 38/42 (90.47%) of the patients had OAEs present in their right ears. On the contrary, OAEs were absent in 6/42 (14.28%) of the right ears and 4/42 (9.52%) of the left ears. A study of cochlear microphonics was undertaken and it was observed that all (100%) of the patients had absent cochlear microphonics in both ears and a reverse polarity was also analyzed. Auditory brain stem response (ABSR) in the study population showed that all the patients showed threshold levels above 110 dB in all the left ears with prolonged latency periods. One patient had ABSR values in the right ear. MRI studies of all the patients were undertaken in this study and found that there was no inner ear abnormality noticed in all the patients (100%). Of the 42 patients studied, 37 (88.09%) of them were found eligible for amplification. In view of absent cochlear microphonics, cochlear implantation was not attempted in any of them. Rest of them 5/42 (11.90%) were comfortable without any rehabilitation options. Of 37/42 patients (88.09%), none of them found any significant benefit with amplification. Speech strategy was advised for them.

DISCUSSION

In the above descriptive study conducted at the Department of ENT, Government Medical College, Kozhikode, 42 patients with ANSD were analyzed for their symptoms, audiological evaluation, and radiological evaluation. The sociodemographic, clinical, and audiological

Table 6: The incidence of various risk factors in the study group (*n*=42)

Other etiological factors	Number	Percentage	Absent	Percentage
Ototoxic drugs	3	7.14	39	92.85
Premature birth	10	23.80	32	76.19
Low birth weight	23 54.76% <1.5 kg–02 >1.5kg–21	4.76 50	19	45.23
NICU admission	10	23.80	32	76.19
Consanguinity	5	11.90	37	88.90
Peripheral neuropathy	4	9.52	38	90.47
Neurodegenerative disease	1	2.38	39	92.85

NICU: Neonatal intensive care unit

Table 7: The pure tone average values among the study group in their right ears (*n*=42)

Pure tone average	Number (%)
Minimal (16–25 dB)	5 (11.9)
Mild (26–40 dB)	6 (14.28)
Moderate (41–55 dB)	22 (52.38)
Moderate-to-severe (56–70 dB)	3 (7.14)
Severe (71–90 dB)	4 (9.52)
Profound (above 90 dB)	2 (4.16)

Table 8: The pure tone average values among the study group in their left ears (*n*=42)

Pure tone average	Number (%)
Minimal (16–25 dB)	4 (9.52)
Mild (26–40 dB)	14 (33.33)
Moderate (41–55 dB)	14 (33.33)
Moderate-to-severe (56–70 dB)	4 (9.52)
Severe (71–90 dB)	4 (9.52)
Profound (above 90 dB)	0 (0)

data were studied to determine the audiological profile of ANSD with an overview of etiology. Further, this study also assessed the various rehabilitation strategies available and the patient benefit with them. Among the study group, a total of 42 patients, majority were in the age group of <20 years (80.95%). This was consistent with the study of Berlin *et al*.,[4] where 153 of 260 patients studied fell in the age group of <18 years. Of these, 69% were female and 31% were male, this was different from the study of Berlin *et al*. where the majority were males which were not considered significant in this study. This difference could be attributed to the large sample of the study by Berlin *et al*. (260 patients). All of them presented with hearing problems. Some had difficulty in understanding while others had difficulty in hearing. Majority of them (97.6%) had bilateral hearing problem and only one of them presented with unilateral hearing problem (left side). This was again consistent with the study of Berlin *et al*.[4] where 92.69% had bilateral disease, whereas 07.31% had unilateral disease with the left side predominance (68.42%). While 14.3% of them had vertigo, 28.6% of them complained of tinnitus (4.8% involving the right ear alone). None of them found it debilitating in contrast with the study by Prabhu and Jamuar[13] where majority had moderate degree of functional impairment due to tinnitus. The study also concluded that no significant association between onset of hearing loss and tinnitus could be found. One of the characteristic observations was that all of the study participants had difficulty in speech discrimination, more aggravated in the background of noise. The severity of this varied, with one end showing speech discrimination abilities compatible with day-to-day activities and at the other end, we had people who presented with mask-like faces, completely oblivious of their environment. This observation was found to be similar to those of Rance[5] where speech perception ability of the ANSD group was found to be poorer than the matched normal and sensorineural group. In spite of poor speech discrimination, 59.5% had good clarity in speech. Of the 42 patients studied, 54.8% had a history of low birth weight, 33.3% had preceding viral infections, and 23.8% had prematurity and NICU admissions. Less than 15% of them had a history of consanguinity, head trauma, peripheral neuropathy, and ototoxic drug exposure and neurodegenerative diseases. Due to the multiple risk factors in many of them, there was considerable overlap in percentages. In 30.95%, no identifiable cause could be found. In the study by Berlin *et al*.,[4] the major risk factors were hyperbilirubinemia (48%) and premature birth (47%), with only 7% having history of low birth weight. As genetic study was not done, the causation could not be attributed to these risk factors alone in the present study. Further, probing into the genetic workup might be needed for the same. In the category of low birth weight, 4.8% of patients were <1.5 kg with the lowest being 750 g. However, no significant association was found between birth weight and age of onset of symptoms. In a case report by Salvinelli *et al*.,[14] preserved OAEs were seen in post parotitis patients in the presence of profound SNHL and abnormal ABR. It was attributed to the tropism of virus to the inner hair cells or sparing of outer hair cells due to difference in antigenicity. In our study, of the 33.3% having viral infection preceding the onset of symptoms, 50% had viral parotitis. However, contrary to the presentation in the report, all of them in our study had bilateral hearing problem. Although 23.8% of them had prematurity and NICU admission ranging from 10 to 28 days, no significant association was found with the onset of disease. Similar to the study by Unal and Vayisoglu,[10] consanguinity, ototoxicity, and head trauma were also present in few people, with a history of consanguinity and head trauma in 11.9% and ototoxicity in 7.1%. On examination, 9.5% of them had peripheral neuropathy and one of them had neurodegenerative disease in the form of hereditary spastic paraplegia. The presence of Grade I bilateral retraction of tympanic membrane in 21.4% of them was not found to be statistically significant. Majority (78.6%) had normal external auditory canal and tympanic membrane findings. In pure tone audiogram, majority fell in the range of mild-to-moderate hearing loss with low-frequency type. This was consistent with the findings of Berlin *et al*.[4] study, where 16.28% had mild-to-moderate hearing loss. A clinching factor here was poorly correlating PTA and speech audiometry. Speech audiometry becomes a reliable pointer here. Most of the time patients presented with difficulty in comprehension or poor academic performances. The discrepancy between the PTA and speech audiometry prompted us to probe deeper, thus unveiling the ANSD. A study conducted by Sinha *et al*.[15] focuses on the importance of doing speech audiometry routinely by

highlighting a case of coexisting ANSD and conductive hearing loss, which without speech audiometry would have been managed as a case of otosclerosis. About 90.5% had speech reception threshold more than 45, implying the need for rehabilitation options. About 92.9% had normal tympanogram. The changes in tympanogram did not influence the disease status. All of them had absent acoustic reflexes. In the earlier days, the diagnostic factor in ANSD was the presence of OAEs and cochlear microphonics. However, as time progressed, it was found that the OAEs and cochlear microphonics need not always be present as evidenced by the study of Sharma *et al*.[16] It was noted that in conditions where ANSD coexists with other pathologies (like noise-induced hearing loss), OAE could be absent, and it was also identified that the progressive form of ANSD could damage the outer hair cells. Here, OAEs were present in 85.7% of the right ears and 90.5% of the left ears. In the remaining patients, though no coexisting conditions were noted; all of them had complaints lasting for more than 5 years. Cochlear microphonics (reverse polarity) was found to be absent in all of them – depriving them of cochlear implantation. Reverse polarity was used to prevent the neural artifact. As cochlear response was noted to change polarity when the stimulus is inverted, neural responses do not change polarity. This was shown to uncover AN in the study by Berlin *et al*.,[4] wherein ABR and cochlear microphonics were differentiated based on polarity change. All of them had abnormal ABR in the left ear. In the right ear, only one of them showed normal ABR. This was of the patient who presented with the left unilateral ANSD. MRI of the brain and inner ear detected no significant anomalies. Similar findings were also noted by Starr *et al*.[3] As all of them did not show cochlear microphonics (reverse polarity), cochlear implantation was not tried in this study. Remaining options were amplification and speech strategy. The study by Berlin *et al*.[4] noted that hearing aid had only very minimal benefit in minority of ANSD cases, as the underlying problem of difficulty in speech perception persisted. Moreover, it was argued that amplification modalities may damage the normal functioning outer hair cells, thus doing more harm than good. Many of them opted out of amplification as they were doing well in daily activities without any aids. About 88.1% of them were fitted with hearing aids, but none of them found it beneficial and thus was directed to speech strategies, the result of which needs to be assessed in further follow-up.

CONCLUSIONS

The major risk factor identified in this study for ANSD was low birth weight with prematurity, NICU admissions, and viral infections having significant contributions. On audiological evaluation, hearing loss was of mild-to-moderate range with a low-frequency loss. There was no statistical correlation between pure audiometry values and speech audiometry which was a characteristic observation. OAEs were present in the majority of patients with absent cochlear microphonics (reverse polarity) and acoustic reflexes. Auditory brain stem response was found to be abnormal in all ears with ANSD. After assessing the patient profile, 37 of them were fitted with hearing aids. However, none of them showed any significant benefit, stressing on tailor-made rehabilitation strategies, and further researches into this.

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