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Auditory neuropathy spectrum: the importance of adequate diagnosis

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Abstract

Objective: To evaluate the prevalence of misdiagnosis of the hearing impaired children, in order to investigate the existence of Auditory Neuropathy Spectrum Disorder. **Methods:** Analytical, prospective study conducted with 15 participants (30 ears) aged 10-12 years, with bilateral sensorineural hearing loss, attending a Hearing Rehabilitation Center, accompanied by a multidisciplinary team with otolaryngologist, audiologist, psychologist, teacher and social worker. The participants underwent ENT examination and audiological assessment: tympanometry, acoustic reflexes, otoacoustic emissions test and Auditory Evoked response. **Results:** From the total sample (30 ears), 8 ears (26.7%) presented absent responses in the Auditory Evoked response with the presence of cochlear microphonism. Within the selected eight ears, six (75%) showed presence of otoacoustic emissions test in isolated frequencies and two (25%) ears had otoacoustic emissions test even in the presence of the isolated frequencies. It was found that 26.7% of the ears tested presented results that are compatible with Auditory Neuropathy Spectrum Disorder. **Conclusion:** The results of this study have identified characteristics related to ANSD in 26.7% of ears tested, 6 participants with age over 10 years old, showing that misdiagnosis in hearing loss patients is still very frequent.

Keywords: auditory, child, evoked potentials, hearing loss.

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INTRODUCTION

Hearing impairment leads to changes in the development of auditory and language skills. The development of these skills with focus on auditory information and appropriate speech expression is only possible due to technological devices such as cochlear implants and hearing aids[1].

The current advances in technology and in newborn hearing screening are permitting an earlier diagnosis of hearing loss, improving ENT evaluation and speech therapy focused on speech and language stimulation, aiming the development of auditory skills.

The combination of speech therapy, ENT and audiology interventions added to family support and systematic use of hearing devices provides a better prognosis for hearing impaired children in general[2]. Nonetheless some children, even under ideal conditions of stimulation and support since early childhood, shows auditory and language skills in a level below from what is expected for children of the same age. One possible explanation for this fact is the presence of undiagnosed Auditory Neuropathy Spectrum Disorder (ANSD).

In the last, care assistance and technology available to outpatient services for the diagnosis of hearing loss in children often did not allow to diagnose the ANSD, whereas nowadays newborns can receive an early diagnostic of ANSD due to electroacoustic evaluation methods such as: Otoacoustic Emissions (OAE) and Auditory Brainstem Response (ABR)[3].

The ANSD is a diagnostic used to describe individuals with hearing disorders mainly associated with a dysfunction in inner hair cells (IHC) and auditory nerve synapses and/or auditory nerve only. Unlikely patients with sensorineural hearing loss that present clinical features compatible with impaired function of the inner and outer hair cells, patients with clinical evidence of ANSD have normal function of outer hair cells (OHC)[4].

Individuals with ANSD usually present impaired speech intelligibility and pure tone detection thresholds at levels ranging from normal to severe. It has been seen that the ANSD affects the individual's ability to process quickly changes in the acoustic signal, which means that it affects the auditory temporal process[4].

Some patients with this disorder report the occurrence of fluctuations in hearing, with so-called "good hearing days" and "bad hearing days"[4], besides the complaint of difficulty in understanding speech, especially in the presence of background noise[5].

Objective tests, such as OAE, ABR and acoustic efferent reflexes, evaluate the function of peripheral and central auditory pathways. Although measures of auditory behavior of central function provide general information on communication performance, objective measures show high sensitivity in neural testing[6].

The ANSD is characterized by hearing loss with absent or severely altered ABR, normal function of OHC obtained by present OAE and the presence of cochlear microphonism (CM)[7]. Through tests such as OAE and ABR, it is possible to identify if the eighth cranial nerve (vestibulocochlear) function is altered, considering that in ANSD cases the OHC function may be normal[5].

The increasing number of newborn hearing screening programs that uses methods such as OAE and ABR is significantly contributing to the diagnostic of neural hearing loss in newborns and young infants. From this population, children that were screened and presented abnormal ABR and presence of OAE were the first ones where the presence of a neural hearing impairment was suspected[8].

Speech perception abilities reduce with an increase in the degree of hearing loss. The amplification given by hearing aids is used to improve the loss of sensitivity and it usually results in good outcomes to speech perception. In contrast, a dysfunction of the auditory nerve may not impact auditory sensitivity significantly, but it usually jeopardizes speech understanding, which is generally poor in these cases. This is because the auditory nerve is the main pathway to the transmission of neural code for speech. In general, amplification may not improve understanding of speech when the auditory nerve is compromized[8,9].

Aim of Study

To evaluate hearing impaired children, aging from 10 to 12 years old, who are hearing aids users, in order to investigate the existence of Auditory Neuropathy Spectrum Disorder (ANSD).

MATERIALS AND METHODS

It was conducted an analytical and prospective study, with 15 children (30 ears), from a Hearing Rehabilitation Center that were accompanied by a multidisciplinary team, composed by otolaryngologist, audiologist, psychologist, teacher and social worker, in Brasília - DF, in Brazil.

The ages of participants varied between 10 and 12 years. 8 of them (53.3 %) were girls and 7 (46.7 %) were boys. All participants presented bilateral sensorineural hearing loss, from moderate to profound classified as frequency average[10] and were hearing aid users.

A questionnaire[11] was applied with parents of the participants to identify the profile of each participant in terms of prenatal, perinatal and postnatal development, facts related to development of hearing and use of hearing aids.

Procedures performed consisted of: otoscopy using Heine™ otoscope; Tympanometry using MADSEN™ OTOflex 100 - Tympanometer with 226 Hz probe; Distortion product OAEs product using Biologic Scout ™ (diagnostic protocol: 750-8000 Hz 2F1 - F2); ABR (Click to dBnHL 80) with cochlear microphonismresearch (rarefact compared to condensed polarity) with insert earphones and using Bio-logic Navigator Pro diagnostic ABR system™.

Pure tone audiometry data (performed maximum 6 months before the beginning of this study) was extracted from patient file. Middle ear disorders evidenced in tympanometry results were considered as exclusion factor.

The Ethics Institutional Review Committee for Research with Human Beings approved this study (Opinion no. 371/09 SES - DF). Caregivers also signed the informed consent form.

For statistical analysis SPSS version 14.0 was used. Descriptive analysis was performed, comparing mean values, using ANOVA ($P < 0.05$) and correlation between nominal variables with tests: Lambda, Phi and Cramer's V, with significant correlation values greater than 0.5 and $P < 0.05$.

RESULTS

Table 1 presents identification data of participants. It was verified that 66.7% of participants, have an unknown etiology and 66.7% had profound sensorineural hearing loss. The age that caregivers' suspected hearing loss occurred between 15 days to 11 years, the age of diagnosis occurred between 3 months to 11 years, beginning of speech therapy occurred between 3 months and 11 years and the beginning of hearing aid use occurred between 12 months to 11 years.

Participants 5 and 8 are siblings and have the same degree of hearing loss. They belong to a family of three children with hearing loss with late diagnostic, this is due to the inexistence of newborn hearing screening by that time and to the degree of hearing loss that was not as impactful to the development of auditory skills, so it took time to parents to realize that their children have hearing loss.

In Table 2, the results obtained are described in electroacoustic and electrophysiological assessments that were performed.

Figure 1 shows that 26.7% of ears tested (6 participants) had auditory system pre-neural and neural evaluation results, compatible with ANSD with absent ABR and presence of CM.

Figure 2 shows that 8 ears (26.7%) that had absent ABR, had presence of CM. FROM These ears, 6 (75%) showed presence of OAE in isolated frequencies and 2 ears (25%) absence in OAE.

Figure 3 shows the result of OAE test, in the ears that had presence of OAE in isolated frequencies, but with "fail" in the final test result.

In Figure 4 there is a correlation between the duration of use of hearing aids and ears that show presence of OAE in single frequencies in children with characteristics of ANSD.

Table 1. Distribution of the sample according to age, gender, etiology, degree of hearing loss, age that parents suspected the hearing loss, age of diagnosis, beginning of speech therapy and beginning of hearing aids use.

Participant	Age	Gender	Etiology	Degree of HL	Age when HL was suspected	Age of diagnostic	Beginning of Speech Therapy	Beginning of HA Use
1	10y	Male	Rubella	Profound	8m	8m	1y 6m	1y
2	11y	Female	Idiopathic	Profound	2y 3m	2y 3m	2y 5m	2y 11m
3	12y	Male	Idiopathic	Severe	15 days	3m	3m	3y
4	12y	Female	Toxoplasmosis	Severe	1y	1y 8m	2y	2y 9m
5	10y	Male	Idiopathic	Moderate	10y	10y	10y	10y
6	10y	Male	Idiopathic	Profound	7m	9m	1y	1y 3m
7	12y	Male	Rubella	Profound	8m	2y	2y 3m	2y 3m
8	12y	Female	Idiopathic	Moderate	11y	11y	11y	11y
9	10y	Female	Rubella	Profound	1y	1y 4m	1y 4m	1y 10m
10	11y	Male	Consanguinity	Profound	8m	11m	11m	1y
11	10y	Female	Idiopathic	Profound	2m	8m	1y 4m	1y 5m
12	11y	Male	Idiopathic	Profound	3m	1y	4y	2y 6m
13	11y	Female	Idiopathic	Severe	2a	2y	8y	5y
14	11y	Female	Idiopathic	Profound	6m	1y 3m	2y	1y 6m
15	10y	Female	Idiopathic	Profound	Parents do not remember	8m	1y	1y

y: year; m: month; HL: Hearing loss; HA: Hearing aids.

Table 2. Electroacoustic and Electrophysiological Evaluation.

Participant	OAE		ABR		Cochlear Microphonism		ANSD Characteristics	
	Right Ear	Left Ear	Right Ear	Left Ear	Right Ear	Left Ear	Right Ear	Left Ear
1	Absent	Absent	Absent	Absent	Absent	Absent	No	No
2	Absent	Absent	Absent	Absent	Absent	Absent	No	No
3	Present in 8.0 Hz	Absent	Absent	Absent	Present	Absent	Yes	No
4	Absent	Absent	Present Waves I, III, V	Present Waves I, III, V	Absent	Absent	No	No
5	Absent	Absent	Present Waves I, III, V	Present Waves I, III, V	Absent	Absent	No	No
6	Absent	Present in 4.0 Hz	Absent	Absent	Present	Present	Yes	Yes
7	Absent	Present in 3.0 Hz	Absent	Absent	Present	Present	Yes	Yes
8	Absent	Absent	Present Waves I, III, V	Present Waves I, III, V	Absent	Absent	No	No
9	Absent	Absent	Absent	Absent	Absent	Absent	No	No
10	Present in 3.0 Hz	Absent	Absent	Absent	Present	Absent	Yes	No
11	Absent	Absent	Absent	Absent	Absent	Absent	No	No
12	Absent	Present in 2.0 and 3.0 Hz	Absent	Absent	Absent	Present	No	Yes
13	Absent	Absent	Present Wave I, III, V	Present Wave I, III, V	Absent	Absent	No	No
14	Absent	Absent	Absent	Absent	Absent	Absent	No	No
15	Absent	Present in 2.0 Hz	Absent	Absent	Absent	Present	No	Yes
			TOTAL				4	4

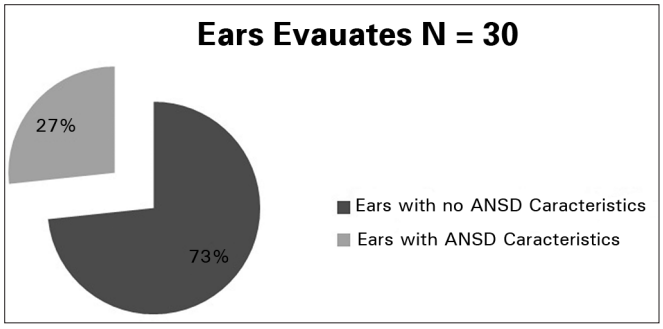

Figure 1. Ears compatibles with ANSD.

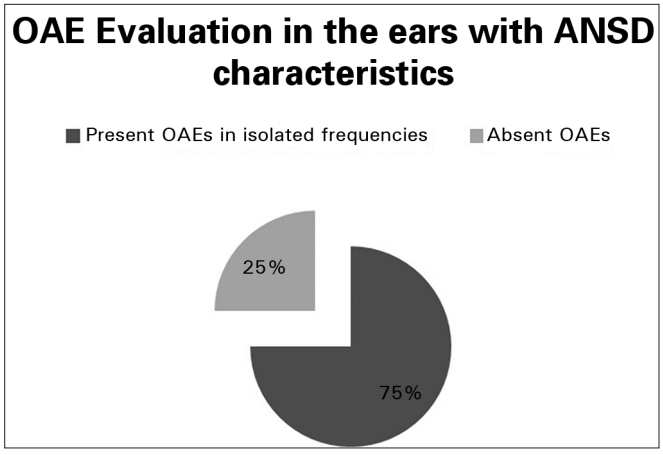

Figure 2. OAE evaluation of the ears that showed ANSD characteristics.

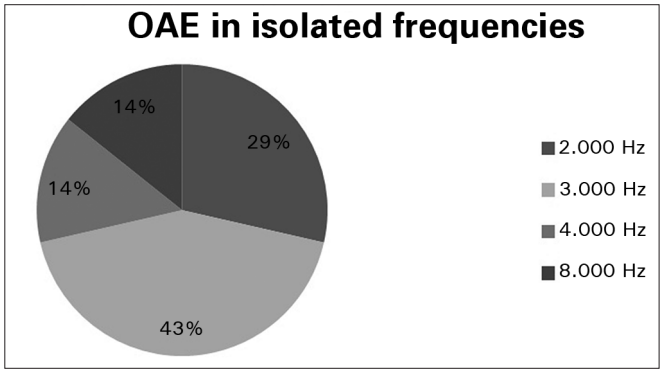

Figure 3. Distribution of OAE presence according the frequency of response.

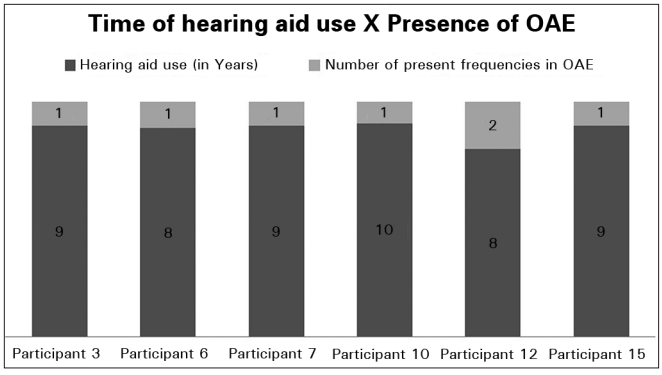

Figure 4. Time of hearing aid use X Presence of OAE.

DISCUSSION

The diagnosis of hearing loss in participants of this study occurred between 3 months to 11 years old. Similar data was observed in a study[7] where 218 deaf children were evaluated and, in most cases, were diagnosed between 11 months to 12 years of age (mean age 4 years). This results shows that the diagnosis of hearing loss is still happening late, even with the advances in technology for diagnostic methods.

Regarding the degree of hearing loss, this study found that 66.7 % of the sample had profound hearing loss, 20% severe and 13.3% moderate hearing loss. Similar results were found in study[7] where 86.69% of participants had profound hearing loss, 10.55% with severe and 2.75 % with moderate to severe hearing loss. In the literature, other authors agree that there is a higher prevalence of profound hearing loss.

The electrophysiological measures of auditory function had a great importance in order to understand the alterations that were found in this study. The results obtained from the tests performed showed characteristics described as ANSD, such as: Present OAEs; Absent ABR with the presence of CM with the inversion the polarity of the click. It was observed that 26.7 % of ears evaluated (6 participants) presented characteristics of ANSD in the pre-neural and neural auditory system assessment, revealing absent ABR with the presence of CM. From these ears, 75% showed present OAEs in isolated frequencies, however, without reaching the criterion "pass" in the test.

Despite the plastic tube of insert phones were not tied up during the ABR test in order to confirm the presence of the CM[5], OAE test was performed. In this test cochlear function responses were obtained in some isolated frequencies, what maintains the reliability of the CM research, which is also a response from the cochlear function.

The features found in physiological tests in this study were consistent with results found in literature. In a study where 10 patients were evaluated in order to find the presence of ANSD, 8 showed evidence of auditory neuropathy. This disorder is characterized by normal function of OHC (Presence of OAE and CM) and abnormal function of the auditory pathways beginning in the eighth cranial nerve (absent or severely abnormal ABR).

Another study with a 59 participant sample[12], three of them were diagnosed with auditory neuropathy by presenting present OAEs, absent ABR and extended CM. Four other participants had absent OAEs and present ABR, although, when the polarity of stimulus were inverted it was observed a mirror image, which is compatible with CM.

Newborn hearing screening was not performed with participants of this study by the time they were born, since there was no implementation of universal newborn hearing screening programs in the hospitals where they were born. There is therefore no way to know whether or not these children would have the present OAE in all frequencies, indicating integrity of the cochlear function (OHC). What showed evident in the results of OAEs in this study were OAE responses in isolated frequencies. It is known that OAEs may be present at birth, but may fade over time. Thus, the absence of OAEs or presence in isolated frequencies does not exclude the ANSD diagnosis[13].

Some patients with ANSD have Absent OAEs, but show signs of hair cells function that are evident in the CM, which is an electric evoked potential of hair cells that can be recorded in a non-invasive way with the ABR test[8].

In the present study, it was identified responses in isolated frequencies in OAE test, this occurred in 2.0, 3.0, 4.0 and 8.0 kHz. In other studies there is no information whether this loss of cochlear function that happens in ANSD begins at high frequencies or low frequencies, or even if it occurs randomly. The data described in a study[14] shows that DP OAEs has been detected in 98.1% of participants with ANSD 3 to 6 years after diagnosis and that the amplitude of OAEs was reduced by less than 3 dB in 77.3% of the sample and by more than 3 dB 20.8% of the sample.

All participants who presented characteristics of the ANSD in this study were fitted with hearing aids, and showed present OAEs in isolated frequencies. The same was observed in another study[14] where 18 ears (11.7%) with ANSD fitted with hearing aids. The results showed that the decrease in the amplitude of the OAE in the ears fitted with hearing aids was higher than the non-aided ears. These results emphasize the possible role of amplification in the reduction of otoacoustic emissions in ANSD[14].

CONCLUSION

The results of this study have identified characteristics related to ANSD in 26.7% of ears tested, 6 participants with age over 10 years old.

This study leads to a reflection on the lack of early diagnosis in ANSD that may have hindered the adequate intervention with hearing aids, hearing tests, and specific therapeutic monitoring in this population. Other studies with larger samples should be conducted for a better investigation of ANSD.

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