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Speech perception in users of hearing aid with auditory neuropathy spectrum disorder

Percepção de fala em deficientes auditivos pré-linguais com desordem do espectro da neuropatia auditiva usuários de aparelho auditivo de amplificação sonora

ABSTRACT

Purpose: To analyze speech perception in children with pre-lingual hearing loss with auditory neuropathy spectrum disorder users of bilateral hearing aid. **Methods:** This is a descriptive and exploratory study carried out at the Research Center Audiological (HRAC/USP). The study included four children aged between 8 years and 3 months and 12 years and 2 months. Lists of monosyllabic words, two syllables, nonsense words and sentences, the Infant Toddler–Meaningful Auditory Integration Scale (IT–MAIS) and the Meaningful Use of Speech Scale (MUSS), hearing, and language categories were used. All lists were applied in acoustic booth, with speakers, in free field, in silence. **Results:** The results showed an average 69.5% for the list of monosyllabic words, 87.75% for the list of two-syllable words, 89.92% for the list of nonsense syllables, and 92.5% for the list of sentences. **Conclusion:** The therapeutic process that includes the use of bilateral hearing aid was extremely satisfactory, since it allowed the maximum development of auditory skills.

RESUMO

Objetivo: Analisar a percepção de fala em crianças portadoras de deficiência auditiva pré-lingual com desordem do espectro da neuropatia auditiva (DENA) usuárias de aparelho de amplificação sonora individual (AASI) bilateral. **Métodos:** Trata-se de um estudo descritivo e exploratório realizado no Centro de Pesquisas Audiológicas da Sessão de Implante Coclear do Hospital de Reabilitação de Anomalias Craniofaciais (HRAC/USP). Foram avaliadas 4 crianças com idade variando entre 8 anos e 3 meses e 12 anos e 2 meses. Foram utilizados: listas de palavras monossílabas, dissílabas, palavras sem sentido e sentenças, Escala de Integração Auditiva Significativa para Crianças Pequenas (IT-MAIS) e Questionário de Avaliação da Linguagem Oral (MUSS), categorias de linguagem e audição. Todas as listas foram aplicadas em cabine acústica, à viva-voz, em campo livre, no silêncio. **Resultados:** Os resultados apresentaram média de 69,5% para a lista de palavras monossílabas, 87,75% para a lista de palavras dissílabas, 89,92% para a lista de sílabas sem sentido e 92,5% para a lista de sentenças. **Conclusão:** O processo terapêutico aplicado, que incluiu o uso do AASI bilateral, foi extremamente satisfatório, uma vez que possibilitou o desenvolvimento máximo das habilidades auditivas.

Study carried out at the Audiological Research Center, Craniofacial Anomalies Rehabilitation Hospital, Universidade de São Paulo – USP – Bauru (SP), Brazil.

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INTRODUCTION

Hearing and communication skills are distinct features of the human existence, considered the greatest contributors for the well-being of any individual. However, the auditory sensory deprivation affects the development of the hearing function and the oral language, and may result in emotional, educational, social, and cultural implications⁽¹⁾.

The auditory neuropathy spectrum disorder (ANSD) is a dysfunction of the synapses of inner hair cells and/or auditory nerve. The individual with ANSD presents the functioning of the outer hair cells, however, the function of the vestibular-cochlear nerve is altered⁽²⁾. Clinical characteristics include from auditory thresholds within normality to severe and/or profound bilateral hearing loss⁽²⁻⁴⁾. It is important to highlight that the ANSD may be observed in individuals of all ages⁽⁵⁾ and that there are many possibilities for the exact location of the alteration, the ANSD may be caused by failure of inner hair cells and/or the synaptic junction between these cells and the fibers of the VIII cranial pair, and/or the fibers of the VIII cranial pair themselves, and/or in the biochemical basis and release of neurotransmitters, or even a combination of the structures mentioned⁽²⁾.

The causes of this alteration may be associated to family history, the permanence in neonatal intensive care unit, hyperbilirubinemia, premature birth and neo and perinatal asphyxia, or even syndromes, such as, the Charcot-Marie-Tooth syndrome, Friedreich's ataxia, and the Mohr-Tranebjærg syndrome. Thus, it is important to consider that an early and accurate diagnosis is essential for these patients^(2,3).

In the infant population, clinical characteristics of ANSD may determine additional difficulties for the development of hearing abilities. This way, the hearing habilitation and rehabilitation of children with ANSD are a challenge, because, in this population the neural function alteration may or may not lead to impairment of hearing perception; in the cases when these difficulties are diagnosed, its biopsychosocial development may be affected.

Due to this complexity of the ANSD condition, different treatment options have been considered and indicated. When an intervention by means of hearing aids is needed, the resources extend from the use of an individual hearing aid (IHA) to frequency-modulated system (FM system) and cochlear implant (CI).

As for the use of the IHA by this population, when indicated as a rehabilitation strategy, it stimulates the residual hearing and may improve the hearing thresholds⁽⁶⁾. Some authors reported that the IHA may contribute to a better hearing detection of the sounds^(3,6); however, there may not necessarily be an improvement in auditory discrimination⁽⁷⁾.

In a study⁽⁸⁾ with children with ANSD users of IHA, the authors found a significant improvement in the hearing condition, both for the results of the tests of speech perception carried out in silent and for the audiometric evaluation in free field with research of the functional gain among children with ANSD.

Another study⁽⁹⁾ showed that children with ANSD users of IHA were able to reach global speech comparable to the results of speech perception of children with ANSD users of CI.

This way, what is noticed with the survey of the studies carried out with the population of children with ANSD users of IHA is that the results related to the hearing performance and the benefits by means of use of this device are widely variable. Besides, there are few scientific evidences related to the evaluation of speech recognition of children with ANSD users of IHA. Thus, this research had the objective of analyzing the speech perception of children with pre-lingual hearing impairment with ANSD users of bilateral IHA.

METHODS

It is a descriptive and exploratory study carried out in the Audiological Research Center of the Cochlear Implant Department of the Craniofacial Anomalies Rehabilitation Hospital (HRAC/USP), in the city of Bauru (SP), Brazil.

The project was approved by the Audiological Research Ethics Committee of the Cochlear Implant Department of the HRAC/USP, process No. 199/2011. All subjects who took part in this study signed the informed consent.

For the sample, some inclusion criteria were adopted:

- children aged under 18 years old, diagnosed with ANSD, verified by the absence or partial presence of answers in the research of the brainstem evoked auditory potential (BEAP), presence of otoacoustic emissions mentioned are identified in the first evaluation performed and/or cochlear microphonic presence and absence of acoustic reflexes;
- slight-to-moderate degree of hearing loss;
- users of bilateral IHA.

Subjects who did not take the speech perception test and subjects who did not agree in taking part in this research were excluded from the study.

This way, the present study consisted of a sample of four children users of IHA, enrolled at the Audiological Research Center of the Cochlear Implant Department of the HRAC/USP. The age of the sample varied between 8 years and 3 months old and 12 years and 2 months old, and the time of use of the IHA varied between 5 years and 8 months and 9 years and 3 months. All participants used the IHA daily for, at least, 9 hours.

In Chart 1, there is the characterization of the participants regarding gender, risk factors, age of the audiological intervention, time of use of the IHA, and age of evaluation.

The Chart 2 follows the description of the cases related to the information of the audiological tests.

The procedures for the evaluation of speech perception were as follows: list of recognition of monosyllabic and disyllabic words; list of recognition of nonsense syllables; and list of recognition of sentences⁽¹⁰⁾. All lists were applied in a free-field system of acoustic booth, in silent condition.

A retrospective analysis of the data in the chart of the participants was performed regarding the Infant Toddler-Meaningful Auditory Integration Scale (IT-MAIS)⁽¹¹⁾, Meaningful Use of Speech Scale (MUSS)⁽¹²⁾, and language category, according to the classification suggested by Bevilacqua and Tech⁽¹³⁾, and hearing category, according to the proposal of classification of development of hearing skills suggested by Geers⁽¹⁴⁾. A research

of the tone audiometric thresholds in free field with hearing aid devices was also performed.

Previously, the evaluator clarified the possible doubts of the participants, without interfering at all in their responses. It is noteworthy that all participants carried out the evaluation of the speech perception without the aid of lip reading and with the use of the bilateral IHA.

The results were expressed in percentage of correct answers, which varied from 0% to 100%. The higher the score obtained in the test, the better the performance was considered.

RESULTS

The individual results obtained in the application of the speech perception test are represented in Table 1.

Table 2 shows the data of children with ANSD users of IHA obtained in the use of the speech perception test, related to the following aspects: mean, standard deviation, maximum, and minimum.

It was observed that, in relation to the evaluation of parents as for the hearing development of the child, evaluated by the IT-MAIS questionnaire, before the intervention the results varied from 0 to 2.5%. After the use of the IHA, the results varied from 60 (1 year of use of the IHA) to 100% (3 years of use of the IHA). As for the MUSS questionnaire, the results before the intervention varied from 0 to 1.5%; after the use of the IHA, the results varied from 40 (1 year of use of the IHA) to 100% (3 years of use of the IHA).

All children in this study presented the recognition of words in an open set, compatible with the hearing category 6⁽¹⁴⁾, and are fluent in oral language, presenting the development of language skills according to the category 5⁽¹³⁾. It is important to highlight that all children in this study were enrolled in speech language and audiology therapy that focus only on their school difficulties.

The tonal audiometric thresholds in free field, presented by the participants of this study with the use of hearing aids, may be observed in Table 3.

Chart 1. Characterization of the sample

Subjects	Gender	Risk factor	Age of audiological intervention	Time of use of the IHA	Age at the evaluation
1	Male	Hyperbilirubinemia	2 years and 3 months	7 years and 10 months	10 years and 1 month
2	Male	Prematurity/meningitis	6 months	8 years and 6 months	9 years
3	Male	Prematurity/hyperbilirubinemia	2 years and 7 months	5 years and 8 months	8 years and 3 months
4	Male	Prematurity/permanence in the ICU	2 years and 11 months	9 years and 3 months	12 years and 2 months

Caption: IHA = individual hearing aid, ICU = intensive care unit

Chart 2. Description of the audiological findings of the sample

Subjects	Timpanometry	Reflex (contra and ipsilateral)	Hearing loss degree	BEAP (presence or absence)	T-OAE	DP-OAE
1	Type A	Absent	Mild bilateral hearing loss	Absent with the presence of cochlear microphonism bilateral	Absent in all bilateral frequencies	Absent in all bilateral frequencies
2	Type A	Absent	Mild and severe bilateral hearing loss	Absent with the presence of cochlear microphonism bilateral	Present in all bilateral frequencies	-
3	Type A	Absent	Mild bilateral hearing loss	Absent with the presence of cochlear microphonism bilateral	Absent in all bilateral frequencies	RE: Absent in all frequencies LE: Present in the frequency of 2.5 kHz
4	Type A	Absent	Mild bilateral hearing loss	Absent with the presence of cochlear microphonism bilateral	Absent in all bilateral frequencies	Absent in all bilateral frequencies

Caption: BEAP = brainstem evoked auditory potential; T-OAE = transient otoacoustic emissions; DP-OAE = distortion product otoacoustic emissions; RE = right ear; LE = left ear

Table 1. Individual values obtained in the use of speech perception test of children with auditory neuropathy spectrum disorder users of bilateral hearing aid

Subjects	Speech perception test			
	Case study (n=4)			
	Monosyllables	Disyllables	Nonsense syllables	Sentences
	%	%	%	%
1	70.00	76.00	60.00	72.00
2	80.00	88.00	84.00	99.00
3	92.00	96.00	73.00	98.00
4	92.00	94.00	84.00	100.00

Table 2. Results obtained in the use of the speech perception test among children with auditory neuropathy users of hearing aid

	Speech perception test			
	Case study (n=4)			
	Monosyllables	Disyllables	Nonsense syllables	Sentences
	%	%	%	%
Mean	69.50	87.80	89.90	92.50
Standard deviation	11.50	8.90	11.40	13.50
Minimum	70.00	76.00	60.00	72.00
Maximum	96.00	94.00	84.00	100.00

Table 3. Tonal thresholds in free field with hearing aid of the evaluated subjects

Individuals	Frequencies					
	250 Hz	500 Hz	1 kHz	2 kHz	3 kHz	4 kHz
1	20 dB	20 dB	20 dB	20 dB	45 dB	45 dB
2	20 dB	20 dB	20 dB	20 dB	30 dB	30 dB
3	20 dB	30 dB	35 dB	35 dB	40 dB	40 dB
4	20 dB	20 dB	35 dB	35 dB	40 dB	40 dB

DISCUSSION

The findings of this study allowed the observation of individuals with ANSD users of bilateral IHA, participants of this study presented high performance rates in the tests of speech perception performed.

When verified in the literatures what the authors reported about the indication of IH for the habilitation and rehabilitation among ANSD cases, it is possible to notice that there are different positions of scholars regarding this indication; that is, some reported that the development of hearing abilities by means of use of the IHA among children with ANSD is not reached out properly and effectively and, therefore, the indication of CI is the best option for treatment of these cases. However, other scholars reported that the development of hearing skills being possible, exclusively by means of use of the IHA among children with ANSD.

According to Hood⁽⁵⁾, some adult individuals with ANSD reported little or even no benefit with the use of IHA. Other authors also reported that the use of IHA among subjects with ANSD did not report improvement in the performance⁽¹⁵⁾. However, studies^(16,8) reported that the use of IHA among subjects with ANSD improved speech recognition in up to 30%.

In this study, the results showed that all children presented satisfactory results with the use of IHA, considering that the values presented estimated average of 69.5% for the list of monosyllabic words, 87.75% for the list of disyllabic words, 89.92% for the list of nonsense syllables, and 92.5% for the list of sentences. These results showed that the children evaluated are able to aurally recognize words and sentences in an open context, which means without having previous knowledge of what they are listening to.

Besides, it was possible to observe the hearing ability during the evaluation of speech perception, through the instruction of the test given to participants and by reaching category 6 of hearing and category 5 of language. It may also be observed that parents reported good hearing performance, demonstrating

the result of 100% regarding the IT-MAIS and MUSS questionnaires, after 3 years of use of the IHA.

In one study⁽¹⁷⁾, the results demonstrated that there was no statistically significant difference between children with ANSD users of CI, of bilateral IHA, and of IC+IHA. The authors concluded that children with DENA may receive the benefit by means of use of the IHA and present development of their hearing abilities similarly to children with ANSD users of CI.

According to Marino et al.⁽¹⁸⁾, the benefit observed through evaluation tests in free field, of the tests of speech perception or in self-evaluation of the user, obtained from the use of IHA among individuals with ANSD should be considered positive, so that new strategies may be used to improve the speech comprehension and the communication of this individuals.

It is important to highlight that a limitation was found in this study, once that it was not possible to obtain the information about the exact location of the ANSD in the participants, this reinforces the need of investigating the kind of ANSD in this kind of population to help future studies. However, it is valid to consider that three participants did not have the presence of transient-otoacoustic emissions (T-OAEs), indicating that in addition to the neural damage observed by means of the absence of BEAP, there is also sensory alteration, which justifies the use of sound amplification (Chart 2). These information are confirmed by the etiology data described, once that the participants presented risk factors for sensory and neural alterations, suggesting that, in this study, the dysfunction is not only a synaptic or neural alteration.

The investigation of the exact location of the alteration in cases of ANSD may also help, especially, in the indication of resources used in auditory rehabilitation, once that, in the cases in which the alteration is sensory only, it is expected that the individuals have a good evolution of the hearing abilities by the use of IHA; in the cases in which the dysfunction would be sensory and neural the responses of the individuals with the use of IHA or of CI would be unpredictable, but with the use of CI, in this situation, the results would depend on the capacity of the auditory nervous system on dealing with electric stimuli from the device, whether satisfactory or not. Thus, it is important to consider that the accurate diagnosis of the alteration is essential in the population with ANSD.

As for the data regarding age in the audiological intervention, some authors reported that the diagnosis of hearing loss and of the ANSD should be made within the first years of life, once that during this period there is a maturation of the hearing system, important for the good hearing and language development⁽¹⁹⁾. Thus, the results of this study are compatible with the literature.

This way, it was possible to observe that the hearing rehabilitation by use of the IHA, in these cases, allowed improvement in the detection of sounds, auditory discrimination, auditory recognition, and improvement in the percentage of speech perception tests. The children participating in this study reached complex levels of hearing abilities for the conclusion of the evaluation of speech perception in an open set. This result may be attributed to a series of factors, such as the permeability of the therapeutic process, the time of sensory deprivation, the use of IHA and the speech language, and audiology therapy. These aspects contributed and influenced the performance of auditory skills of these children.

This way, the results of the evaluation of speech perception among children with ANSD users of the IHA may help health professionals to reorganize the intervention process, to improve the quality of life of the user.

CONCLUSION

The hearing performance of the participants in this research, whose therapeutic process included the IHA, was extremely satisfactory, once that is allowed the maximum development of the hearing skills.

**NFF was responsible for the elaboration of the research and chronogram, literature review, collection and analysis of the data, writing of the article, submission, and procedures of the article; EHY was responsible for the elaboration of the research, collection, and analysis of the data; MM was responsible for the chronogram, analysis, and writing of the data; OAC was responsible for the correction of the writing of the article and approval of the final version.*

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