CASE REPORT

Auditory and speech development in a 3-year-old boy with auditory neuropathy spectrum disorder

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Received: 27 Nov 2020, Revised: 19 Dec 2020, Accepted: 20 Dec 2020, Published: 15 Apr 2021

Abstract

Background: Auditory neuropathy spectrum disorder (ANSD) in the auditory neural pathway can affect the auditory and speech development of children. Since the symptoms and complications of this disorder are similar in different children, hearing management and rehabilitation can help with better development of speech/language and hearing perception in children with ANSD.

The Case: In this study, the case was a one-year-old boy with ANSD and mild to moderate high-frequency sensorineural hearing loss. He first underwent various audiological examinations. Then, an aural rehabilitation program containing different auditory information and games was provided to him, his family, and caregiver at their home and in the rehabilitation center.

Conclusion: Although auditory neuropathy/dissynchrony in the auditory neural pathway has negative effect on the auditory and speech development, but the children with ANSD can use different inputs for language comprehension and acquisition if they receive effective education, especially auditory training at an earlier age.

Keywords: Auditory neuropathy spectrum disorder; speech perception; auditory training

Citation: Mazaheryazdi M, Akbari M, Abolhasan Choobdar F. Auditory and speech development in a 3-year-old boy with auditory neuropathy spectrum disorder. Aud Vestib Res. 2021;30(2):139-47.

Introduction

Cochlear nerve deficiency is related to auditory neuropathy spectrum disorder (ANSD). Although understanding the cause of ANSD has become challenging, it is possible that the auditory nerve dysfunction and deficits in inner ear’s hair cells and in neurotransmitters associated with otoferlin (OTOF) mutations cause damage in auditory ganglion cells [1]. ANSD can be followed by hyperbilirubinemia, congenital absence or hypoplasia of the auditory nerve, hypoxia, or other impairments that can lead to irregular and synchronous firing of auditory nerve fibers [1,2]. This dyssynchrony in the central auditory pathway can lead to some unusual problems. Since the temporal cues of speech is related to synchrony in auditory pathway, the speech comprehension quality and the integrity of speech signals in children with ANSD are severely impaired, but the frequency and intensity of the process in cochlea are less impaired [2].

The speech perception skills depend on low-
frequency modulations in the presence of background noise or in quiet [3,4]. Apoux et al. indicated that perceived low-frequency spectral information can improve the identification of speech elements [5]. Speech perception of the children with ANSD is significantly different (by 50%) than that of peers with cochlear hearing loss and normal hearing [3,4]. Different strategies have been introduced for improving the speech perception in ANSD children. Cochlear implantation or hearing aids use are two common methods with different benefits for children with ANSD [2,4,6,7].

Severity of damage to neural elements, cochlear frequency resolution, and the function of hair cells and basilar membrane can affect the speech perception. It is also possible that temporal resolution, speech envelope, recordable cortical auditory evoked potentials (CAEPs), poor information about the lesion site, and other related factors can affect the ability of speech intelligibility [6]. Rance et al. reported that CAEPs are correlated with speech perception abilities and amplification benefit [6]. The recording of CAEPs depends on the dendritic potentials that need several milliseconds to drive synchrony; hence, it can be a good index for the speech perception ability [3,4]. The presence of CAEPs and the absence of short-latency evoked potentials indicate that the hearing loss is present and, probably, synchrony in brainstem or axonal action potentials has been destroyed, but it is preserved at the cortical level. Furthermore, when the evoked response potentials (ERPs) are present, there seems to be a sufficient residual synchrony to encode some temporal information needed for speech perception [4,8]. Normal otoacoustic emissions (OAEs) and subsequent normal outer hair cells indicate a good cochlear frequency resolution in some children with ANSD [3]. The frequency resolution depends on peripheral hearing loss and the amount of damage to consonants compared to vowels [4]. One of the important strategies that can improve the conventional speech perception is the enhancement of speech clarity. It emphasis on identification of consonants and enhancement of speech envelope to enhance the salient cues for speech perception [3].

In this case study, we investigated audiologic findings in a 1-year-old boy with auditory neuropathy/dyssynchrony. He was able to recognize the simple words and perceive some simple commands. We performed additional tests on this boy and, at the same time, included him in an auditory training program. We hypothesized that a specified aural rehabilitation program focusing on the frequency discrimination, attention and memory cues, speech clarity, and family support and without using hearing aids can lead to improved conversational speech perception. Since the understanding of the perceptual consequences of auditory neuropathy is challenging especially in very young children, this study presents a rare case of auditory and speech perception in young children with auditory neuropathy.

Case presentation
The case was a 1-year-old boy with symptoms of the auditory neuropathy/dyssynchrony referred to the Auditory Diagnosis and Rehabilitation Center of the School of Rehabilitation, Iran University of Medical Sciences in 2017. His most important complaint was that he had no wave activity in response to the auditory brainstem response (ABR) test, but he could identify and understand their speech and daily conversations. The ABR test was performed in different audiological centers (Fig. 1). All of them recommended cochlear implant. His parents suspected something was wrong with their boy’s hearing, because he could identify speech sounds simple orders, but his electrophysiological test showed no any wave activity; hence, he had been referred to the cochlear implant centers. His parents showed different videos and audio recording of his voice trying to emphasis that their child has no hearing problems. In addition to the use of test battery approach for evaluating his auditory neuropathy according to the protocols in the center, we performed a diagnostic audiological evaluation. The case history of the boy showed that he was born at 34 weeks of gestation. His medical history showed a 20 mg/dL hyperbilirubinemia with a double-volume exchange. The medical staff had made tremendous efforts to survive him. After a few months, the first audiological
evaluation that lasted for four months, showed no any neural activity. The OAE test results reported a healthy cochlea in both ears (Fig. 2).

**Medical assessments**

After recording the case, medical, and otologic histories, a complete ear, nose, and throat examination was performed. Otoscopic examinations were conducted by an otolaryngologist to rule out any external or middle ear pathology that could affect audiometric tests.

**Audiological assessments**

Auditory brainstem response test

We performed ABR test by using the Integrity™ V500 system (Vivosonic Inc., Torento, Canada) for auditory evoked potentials (AEP). We could find wave V at high intensity in the right ear while morphology response showed no integrity in neural activity. We found no wave V in left ear (Fig. 3).

Behavioral assessment

Behavioral observation audiometry (BOA) and visual reinforcement audiometry (VRA) tests were performed at 250 to 4000 Hz by using the

**Fig. 1. Auditory brain stem response with evidence of cochlear microphonic activity in the left (L, 1, 2, and 3 trace) and right ears (R, 5, 7 and 8 trace) with 90 nHL.**

Auditory brainstem response test

Fig. 2. The otoacoustic emissions (red lines) with 60–99% correlation and signal-to-noise ratio > 6 dB in the right ear and 85–96% correlation and signal-to-noise ratio > 6 dB in the left ear.
CA88 audiometer (Pejvak Ava Inc., Iran) with inserting earphone and free-field mode. The results showed mild to moderate high-frequency sensorineural hearing loss that was observed separately in each ear. Immittance audiometry was performed by using the Madsen Zodiac device (Otometrics Co. Denmark) in a standard mode [9]. Tympanometry showed a normal middle-ear function with no acoustic reflexes in any ear. All tests were repeated one month later.

**Speech-language assessment**

Before starting the rehabilitation program, we performed the categories of auditory performance (CAP) scaling to evaluate his auditory perception (Table 1), and the speech intelligibility rating (SIR) test to evaluate his speech production ability (Table 2). The reliability of these two tests for children with hearing loss has already been reported [10-12]. We also used these tests to assess speech perception after their training and rehabilitation. Due to the young age of the boy, we used these two tests as a model and changed some of their items into simple words used in daily conversations.

**Intelligence quotient test**

Nonverbal intelligence quotient (IQ) test was performed by using the Persian version of the Wechsler Intelligence Scale for Children [13,14].

**Cortical auditory evoked potentials assessment**

As the boy grew up and reached the age 2, we evaluated unaided CAEPs by using the HEAR lab system (Frye Electronics Inc., USA) to know whether speech stimuli (sounds) were audible at cortical level. After excluding the results of the recorded random sound, we found out that the measured cortical waveform evoked by each stimulus sound had an acceptable high positive peak (P) of about 200 ms, appropriate to the physiologic age of the boy, for all stimulus sounds of /m/, /g/ and /t/, except for /s/. Since the CAEPs were present at 55 dB SPL for the above mentioned three sounds, it is possible that the behavioral test results indicate a threshold of hearing at normal to mild frequency ranges. Hence, it was possible that there was a mild hearing loss in the reported frequency ranges (Fig. 4).

**Aural rehabilitation program**

Good hearing comprehension and identification of conventional speech using live voice as well as normal hearing in low and mid frequencies.
and hearing loss in high frequency avoided from fitting the hearing aid or offering a cochlear implant. The boy and his family were guided to participate in a special rehabilitation program without using hearing aids or cochlear implant. We tried to enrich his communicative environment by using speech sounds and daily conversations and increase his auditory abilities by using a high-frequency aural rehabilitation. For proper interaction with the boy, we advised his parents, family members, and caregiver to use stimuli sounds, daily conversations and auditory training both in quiet and in the presence of background noise. Also asked them to have daily conversation with higher sound levels along with having a clear and envelope-enhanced speech in quiet and in noisy environment. We scheduled sessions to rehabilitate his auditory attention, auditory memory, and frequency differentiation. To enhance important cues for recognition of speech, we asked him to play different games in quiet and noisy environment. Since previous study has shown that the number of sessions to have effective training is unknown and depends on different situations [15], we did not restrict the training sessions and asked the parents to continue the training for 30 months. We checked all of his audiological evaluation results every two months and where appropriate, additional tests were performed on his hearing behavior. His speech perception was measured by employing five sounds and sometimes by pointing to the members of his doll’s body or his own body under the CAP and SIR tests during the rehabilitation sessions. His speech perception and understanding of speech commands significantly improved.

**Discussion**

Studies have shown that individuals with ANSD are impaired in differentiating signal envelope and temporal fine structure cues from speech signal [3]. Therefore, the enhancement of speech signal’s envelope is important for speech comprehension in individuals with ANSD. Unfortunately, the mechanism of causing difficulty in recognition of speech in the presence of background noise is not clear. It is important to consider the heterogeneity of people with ANSD and the varied etiologies of ANSD in adults or children [16]. They have various speech or language

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<tr>
<th>Category</th>
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<tbody>
<tr>
<td>7</td>
<td>Use of telephone with known listener</td>
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<tr>
<td>6</td>
<td>Understanding of conversation without lip-reading</td>
</tr>
<tr>
<td>5</td>
<td>Understanding of common phrases without lip-reading</td>
</tr>
<tr>
<td>4</td>
<td>Discrimination of some speech sounds without lip-reading</td>
</tr>
<tr>
<td>3</td>
<td>Identification of environmental sounds</td>
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<tr>
<td>2</td>
<td>Response to speech sounds</td>
</tr>
<tr>
<td>1</td>
<td>Awareness of environmental sounds</td>
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<tr>
<td>0</td>
<td>No awareness of environmental sounds</td>
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<table>
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<tr>
<th>Category</th>
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<tbody>
<tr>
<td>5</td>
<td>Connected speech is intelligible to all listeners. Child is understood easily in everyday contexts</td>
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<tr>
<td>4</td>
<td>Connected speech is intelligible to a listener who has a little experience of a deaf person's speech</td>
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<tr>
<td>3</td>
<td>Connected speech is intelligible to a listener who concentrates and lip-reads</td>
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<tr>
<td>2</td>
<td>Connected speech is unintelligible. Intelligible speech is developing in single words when context and lip-reading cues are available</td>
</tr>
<tr>
<td>1</td>
<td>Connected speech is unintelligible. Pre-recognizable words in spoken language. Primary mode of communication may be manual</td>
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Table 1. Categories of auditory performances criteria [11]

Table 2. Speech intelligibility rating categories [10]
test results. Some studies have reported that there is no correlation between pure-tone audiometry results and speech perception performance [12,16]. Some children with ANSD and mild hearing loss can benefit from conventional hearing aids or frequency modulation systems, while the children with significant hearing loss should use cochlear implant [2,7,12]. It can be suggested that an increase in the number of neural elements or an improvement in the neural firing synchrony in the brainstem (perhaps due to the fact that the electric stimuli generated by cochlear implants are presented in the form of discrete pulses, rather than analogue sounds) can improve the speech perception and auditory responses in ANSD children [4]. The important motive to study the boy further was to know whether the aural rehabilitation program can lead to the development of proper speech and language. The boy had mild to moderate high-frequency hearing loss and we consulted with some of our colleagues for fitting hearing aid, but they suggested that, although the increase of amplification can cause good auditory perception in the boy at low frequencies, it can be annoying. However, his appropriate responses to speech stimuli caused his parents to be reluctant to let their child receive hearing aid or cochlear implants. Therefore, with counseling and teamwork, we decided that the boy should participate in an aural rehabilitation program without using a hearing aid or cochlear implant. The rehabilitation strategies and training programs were determined according to the conditions of the boy and his family and based on the total communication approach. The boy, his family, and caregiver were included in the rehabilitation program in an environment with a variety of auditory information and games, conducted at home and in the rehabilitation center. Due to the young age of the boy and his high-frequency hearing loss, we had to use other sensory modalities such as vision for communication; however, our main technique was to use auditory training in the form of an aural rehabilitation program. His progress during training sessions and according to his parents was considerable. The result of present study revealed a significant advantage of speech envelope enhancement by a specific auditory rehabilitation program without using hearing aids or any hearing amplifier device. After some sessions, we evaluated the speech perception by live voice or SIR test and also the auditory performance was evaluated by CAP test. Consistent with the studies that have used the speech sounds /ml/, /g/ and /t/ with spectral emphasis at around 250, 1500 and 3000 Hz, respectively [17], we used them as speech stimuli for testing of speech recognition. It is possible that the open-set speech perception ability vary considerably among children with auditory neuropathy. Some children have appropriate sound recognition ability, while others have poor or no recognition ability [18]. However, as mentioned before, speech perception performance is strongly correlated with the auditory training and

Fig. 4. Auditory cortical responses with different stimuli: /ml/, /g/, /t/ and /s/. Each stimulus demonstrated a fair big single positive peak (p) around 200 ms consistent with the patient’s physiologic age for all stimuli except /s/.
enrichment of educational environment [19]. A study on the mechanism of an effective auditory training program [15] showed that changes in inhibitory neurotransmitters in the central nervous system can increase the latency and decrease temporal precision, which is observed in ANSD children. It seems that auditory training can regulate the function of GABAergic neurons in the central nervous system and increase the levels of inhibitory neurotransmitters. The increase in inhibition can lead to enhanced temporal precision and better speech perception [15,20].

Consistent with the studies that have reported improvements in central auditory processing is related to age, our study showed that speech perception in the presence of background noise develops with the increase of age and is related to the neural processing of the fundamental frequency of the voice (F0) and attention [21]. Some children may enhance auditory processing by perceiving speech in noise as growing up [21,22]. Speech processing and perception in noise depend on different cues as auditory experiences [23]. The neural processing of the F0 is a necessary cue for speech perception and perceptual streaming [21,24]. In this regard, it can be assumed that acceptable thresholds at low frequencies in the boy improved the use of fundamental frequency for speech perception. However, some studies have shown that F0 is not a critical cue for speech perception [25]. Regarding how this boy perceive speech and language appropriately, it can be postulated that the intellectual development level of children with hearing loss is similar or even better than that of norm hearing peers [26]. Verbal/nonverbal perception and the ability to interact with environment can have considerable effects on the cognitive development and various aspects of speech and language. The boy’s high intelligence helped him to perceive different speech stimuli at different situations.

The results showed that the neural synchrony at cortical level was different than the ABR at subcortical level. The peaks detected in the cortical responses indicate slower dendritic activities than in the ABR, depending on synchronous discharge in neural units [27]. Hence, the absence of short-latency potentials in patients with ANSD is not an indicative of speech perception deficits, although it can indicate impaired speech perception in noise. Some studies have pointed out that the development of auditory cortical components P1 and N1 are affected by hearing experiences [6,28]; therefore, it seems that the emergence of cortical waveforms correlated with age is a reason that the boy was still able to hear. The emergence of CAEPs in the boy in our study was consistent with the results of Rance et al. who pointed out that the presence of CAEPs in children with ANSD is related to the speech perception level, not to the degree of hearing loss [6]. Moreover, the P1 robust response in the boy with age-related speech perception indicate that the P1, generated by auditory thalamic and cortical sources, is a biomarker for development of central auditory nervous system [27,29]. Our findings also consistent with the results of Russo et al., who suggested that the auditory stimulation by using an auditory training program may be adequate for development of central auditory pathways in both brainstem and auditory cortex [19]. Overall, our study showed that although the neural synchrony in the auditory nerve pathways plays an important role in speech and language development, it seems that in cases with minor hearing loss, several factors such as IQ, improved high-frequency hearing ability by using an aural rehabilitation, and speech envelope enhancement can affect speech perception in children with ANSD.

Conclusion
Auditory cortex can adjust to different signals in the auditory dyssynchrony. Children with auditory neuropathy spectrum disorder can use different and limited inputs for language comprehension and acquisition if they receive adequate education, especially auditory training at an earlier age.

Acknowledgments
The authors would like to thank the child’s parents for arranging all the appointments and consultations.
Conflict of interest
The authors declare no conflict of interest.

References