Revista Médica del
Hospital General de México

PERMANYER

Check for updates

CLINICAL CASES

Auditory neuropathy spectrum disorder in a patient with normal hearing and its medical management: case presentation and literature review

Jesús A. Silva-Rojas[1,2], Karla L. Ruiz-Lira[1], Emilio Dávalos-González Plata[1], and Pablo A. Ysunza-Rivera[2]*

[1]Audiology and Phoniatrics Service, Unidad 601, Hospital General de México Dr. Eduardo Liceaga, Secretaría de Salud, Mexico City, Mexico; [2]Neuroscience Program, Ian Jackson Craniofacial and Cleft Palate Clinic, Beaumont Health, Royal Oak, MI, USA

Abstract

Auditory neuropathy spectrum disorder (ANSD) is a group of alterations of the auditory system that manifests the presence of otoacoustic emissions and the absence of responses in auditory brainstem response. However, because these patients can have a very diverse clinical presentation, their management is very complex. Patients can present from normal hearing with practically normal development, to profound hearing loss with various comorbidities that require a highly individualized approach. Early detection and close follow-up of ANSD should be a priority to avoid iatrogenic management.

Keywords: *Auditory neuropathy spectrum disorder. Auditory brainstem response. Otoacoustic emissions. Automated auditory brainstem response.*

Introduction

Auditory neuropathy spectrum disorder (ANSD) is thought to be a set of alterations in the auditory system that can include alterations in the synapses of the inner hair cells of the cochlea, or alterations of the spiral ganglion, or myelination defects of auditory nerve axons, or defects in the cochlear nuclei and even a varying degree of involvement of all the aforementioned factors[1]. This spectrum has shown a diagnostic boom from the establishment of early hearing detection programs, such as the performance of otoacoustic emissions (OAE), automated auditory potentials such as neonatal hearing screening tests, and the short-latency auditory provoked potential (SLAEP) test for diagnostic confirmation[2]. Typically, patients with ANSD have present OAEs and absent or seriously altered auditory potentials in all its variants, including SLAEP and PPA, as well as the cochlear microphone response that changes polarity when the polarity of the provocative stimulus changes in the SLAEP[3]. On the other hand, in these patients, the real auditory threshold, or behavioral threshold, can only be determined by behavioral studies such as tonal audiometry in all its variants, including pediatric variants[4]. To reiterate, no electrophysiological study provides reliable hearing threshold answers if ANSD is present. However, audiological studies in a population under 3 years of age must be performed by highly trained, experienced personnel, ideally requiring two specialists for each study/patient, and taking considerable time, and sometimes even several appointments

***Correspondence:**
Jesús A. Silva-Rojas
E-mail: asrmx1@gmail.com

Date of reception: 14-03-2024
Date of acceptance: 13-05-2024
DOI: 10.24875/HGMX.24000021

Available online: 01-04-2025
Rev Med Hosp Gen Mex. 2025;88(2):104-109
www.hospitalgeneral.mx

to reach an audiometric diagnosis. In addition, these studies cannot be reliably performed in children under 6 months of age, and there is a great deal of subjectivity in detecting audiometric thresholds in this age group[5]. A serious problem with this population of pediatric patients with ANSD is that the audiometric thresholds can range from normal hearing to profound hearing loss, likewise, language discrimination can range from normal (100%) to 0%. Evolution is unpredictable and must be monitored very closely with feedback obtained from parents or guardians, and even teachers if necessary[6].

Hearing screening in all cases should ideally be performed with automated potentials (PPA) in both normal pediatric and pediatric populations with high hearing risk. This screening test can not only detect normal hearing or hearing loss in an infant due to cochlear or middle and outer ear damage but also the electrophysiological alteration of the recordings produced by ANSD in the auditory pathway. Unlike OAEs that do not evaluate the auditory pathway and may be normal in ANSD[7]. Because the prevalence/total incidence of ANSD is significantly low in the open population of apparently normal newborns, the use of OAE is widely used to perform neonatal hearing screening, since the possibility of referring a child with ANSD as normal is considered very low[8]. Moreover, the performance of the two tests, SLAEP and OAE, is only indicated in children with high auditory/neurological risk since, as is known, the incidence of this pathology increases considerably in them. Unlike PPAs, SLAEPs are not routinely performed on healthy children because they require specialized personnel for their execution and interpretation, and the time is considerable unlike hearing screening tests, OAEs, and PPAs, which can be obtained in seconds to a few minutes[9].

Patients with ANSD, although with studies of otoacoustic and neurophysiological emissions that are practically the same among all patients, as already mentioned, may present an unpredictable evolution, as well as a very varied clinical picture, and there may be adult patients with difficulty hearing noise, but otherwise normal. Moreover, children with profound hearing loss, multiple comorbidities, and the need for a multidisciplinary approach may even reach cochlear implantation[1,3,9].

ANSD is believed to be multifactorial and can occur in an open population without any apparent neurological or auditory risk factors. On the other hand, it can occur in populations with high audiological or neurological risk, it has even been described as a sequel of Guillain–Barré syndrome or in patients with later-onset neurological diseases such as patients with Freidreich's ataxia[10].

In the open population, ANSD has an unknown frequency in apparently healthy adults. The prevalence of auditory neuropathy varies and can range from 1% to 10% of individuals who have hearing loss, and in newborns with auditory risk factors at birth, the prevalence can reach up to 30%. The prevalence is higher in the pediatric population than in adults[11-14].

Infants with ANSD should not be adapted with hearing aids or cochlear implants due to the absence of responses in SALP and PPA due to the possibility that despite the presence of ANSD they have normal hearing[15]. In these cases, it is suggested that they are frequently cited with the idea of closely monitoring psychomotor development, language development, and auditory behavior reported by the parents[16]. Since, a patient with ANSD without complete studies when simulating profound hearing loss and being adapted with hearing aids runs the risk of presenting irreversible damage at the cochlear level due to exposure to intense sound and consequently losing that valuable function, thus complicating the picture by iatrogenesis. In general, when the parent or guardian is involved, they can provide invaluable information that the specialist can interpret appropriately to improve their diagnosis and individual management according to the evolution of each case[1,6-9].

The purpose of this article is to present a case of a child with ANSD initially diagnosed as hearing loss and with follow-up, who showed electrophysiological improvement *ad integrum* of ANSD.

Case presentation

A female patient born prematurely at 33.4 weeks of gestation with respiratory distress syndrome, pulmonary bronchodysplasia, low birth weight, and anemia of prematurity. There was a risk of sepsis due to a positive maternal history of urine culture for *Enterococcus faecalis*. Placenta previa was identified due to the presence of transvaginal bleeding and a Kerr-type cesarean section was performed at 33.4 weeks of gestation. Apgar from 7 at 1 min and 8 at 5 min. Silvermann of 4. Respiratory whine and thoracoabdominal dissociation treated with CPAP for 72 h did not meet the criteria for asphyxia. She was admitted to the neonatal intensive care unit due to respiratory history and

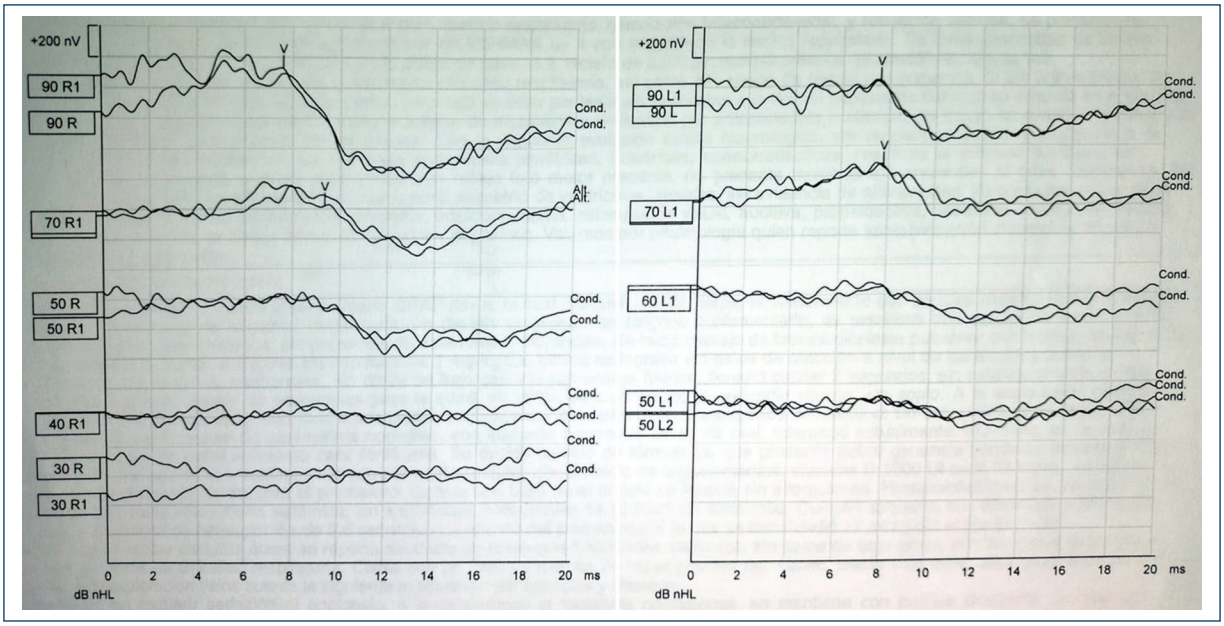


Figure 1. A record of SLAEP was performed at 12 weeks extrauterine, showing poor amplitude, altered morphology, and V-wave with an apparent threshold of 50 dBnHL.

probable sepsis. She received ampicillin, amikacin, fluticasone aerosol, spironolactone, and a blood transfusion. In total, she was hospitalized for 48 days and was discharged. A neonatal hearing screening was performed during her hospital stay, and a normal result was found; however, she was referred to the Audiology and Phoniatrics Service of the Hospital General de México for complementary audiological evaluation due to her history of high hearing risk.

PPALC tests were performed with CE-Chirp stimulus at 3 months of extrauterine life, and SLAEP with V-wave at 50 dBnHL was found, which corresponds to medium-grade hearing loss (Fig. 1). However, when questioning the mother, she refers to an apparent auditory behavior of a hearing norm. For this reason, it was initially diagnosed as middle hearing loss, but the diagnostic protocol for auditory neuropathy was initiated. Again, a month after a month is scheduled to perform SLAEP, this time for cochlear microphone search with stimulus, tone burst with polarity, condensation, and rarefaction (Fig. 2) and to perform OAEs (Fig. 3). There was an inversion of the cochlear microphone with this change in polarity, so probe clamping was performed to rule out the artifact, and normal OAEs were identified. For this reason, ANSD is diagnosed and the mother is instructed to closely monitor auditory behavior, language development, and psychomotor development. Follow-up appointments are given 3 and 6 months later for monitoring of auditory behavior and language development, finding him within normal parameters. A new appointment is made for SLAEP, which is performed at 10 months of age, and V-wave responses are found at normal intensities (better than 30dBnHL) and neurological parameters of the SLAEP within normal limits (Fig. 4). The mother reports auditory behavior of a normal listener, language at the level of a single word, recognizes her relatives. An appointment is made in 6 months and the patient no longer attends. As of the day this article is written, she has not been followed up.

Discussion

As previously mentioned, hearing thresholds in patients with ANSD can vary from normal hearing to profound hearing loss depending on each patient[17], as is the case of this patient since the mother reported normal hearing due to behavioral response even to low-intensity sound stimuli from an early age and this is what has guided us for the successful management of this case. To inexperienced eyes, such a result in SLAEP could simulate hearing loss, the primary management of which would be auditory rehabilitation with hearing aids in both ears; however, it was decided to follow close expectant management. Even in patients already

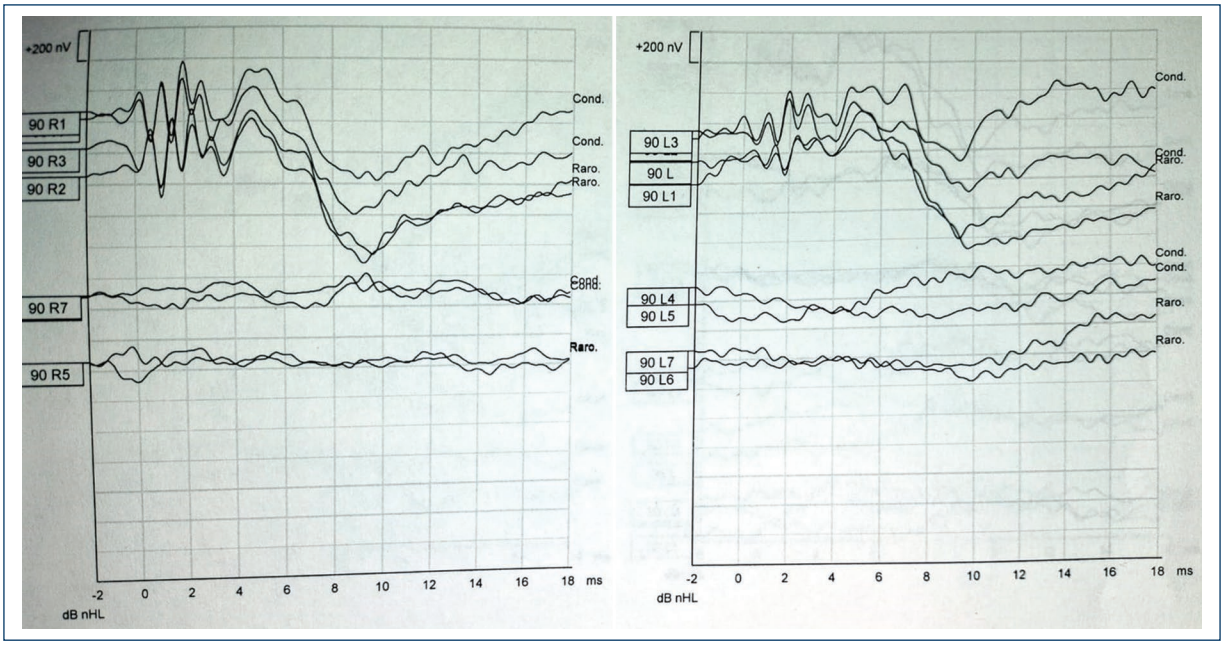
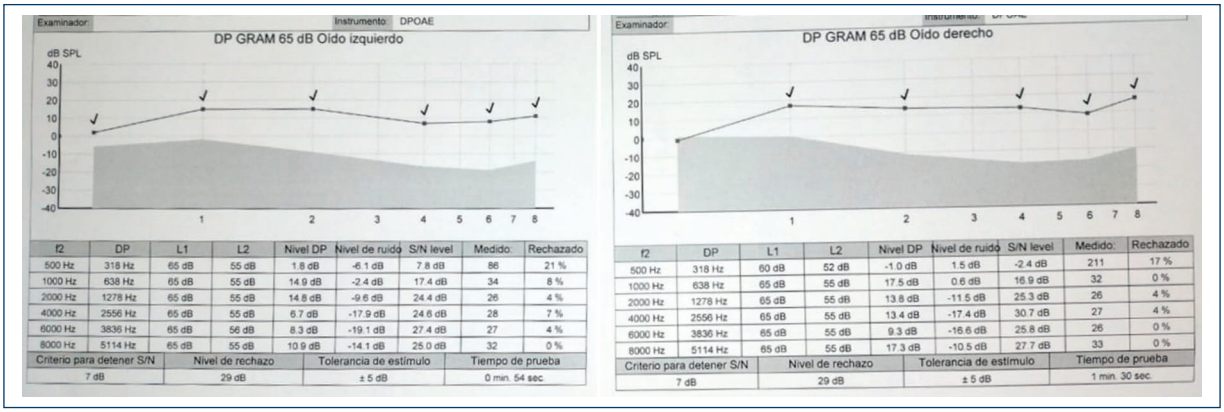


Figure 2. A record of SLAEP for cochlear microphone search was performed at 16 weeks of age with tone-burst, showing the microphone with phase inversion of 180º and below a clamping test where a total absence of responses was observed.



f2	DP	L1	L2	Nivel DP	Nivel de ruido	S/N level	Medido:	Rechazado
500 Hz	318 Hz	65 dB	55 dB	1.8 dB	-6.1 dB	7.8 dB	86	21 %
1000 Hz	638 Hz	65 dB	55 dB	14.9 dB	-2.4 dB	17.4 dB	34	8 %
2000 Hz	1278 Hz	65 dB	55 dB	14.8 dB	-9.6 dB	24.4 dB	26	4 %
4000 Hz	2556 Hz	65 dB	55 dB	6.7 dB	-17.9 dB	24.6 dB	28	7 %
6000 Hz	3836 Hz	65 dB	56 dB	8.3 dB	-19.1 dB	27.4 dB	27	4 %
8000 Hz	5114 Hz	65 dB	55 dB	10.9 dB	-14.1 dB	25.0 dB	32	0 %
Criterio para detener S/N		Nivel de rechazo		Tolerancia de estímulo			Tiempo de prueba	
7 dB		29 dB		± 5 dB			0 min. 54 sec.	

f2	DP	L1	L2	Nivel DP	Nivel de ruido	S/N level	Medido:	Rechazado
500 Hz	318 Hz	60 dB	52 dB	-1.0 dB	1.5 dB	-2.4 dB	211	17 %
1000 Hz	638 Hz	65 dB	55 dB	17.5 dB	0.6 dB	16.9 dB	32	0 %
2000 Hz	1278 Hz	65 dB	55 dB	13.8 dB	-11.5 dB	25.3 dB	26	4 %
4000 Hz	2556 Hz	65 dB	55 dB	13.4 dB	-17.4 dB	30.7 dB	27	4 %
6000 Hz	3836 Hz	65 dB	55 dB	9.3 dB	-16.6 dB	25.8 dB	26	0 %
8000 Hz	5114 Hz	65 dB	55 dB	17.3 dB	-10.5 dB	27.7 dB	33	0 %
Criterio para detener S/N		Nivel de rechazo		Tolerancia de estímulo			Tiempo de prueba	
7 dB		29 dB		± 5 dB			1 min. 30 sec.	

Figure 3. OAE recording of both ears, the schools show normal responses.

diagnosed with ANSD with tonal thresholds by behavioral methods, adaptation is difficult and has to be individualized with each patient[18]. Another rehabilitation route for patients with profound hearing loss is the cochlear implant, and it is very important to know that, in patients with auditory neuropathy, although the behavioral thresholds are of profound hearing loss, expectant management is suggested at least until 2 years of age[19], so if an adequate adaptation is not made. We could generate, as mentioned, irreversible iatrogenic cochlear damage.

Another reason for maintaining expectant management in the auditory rehabilitation of patients with ANSD is because it has been reported that some patients may present recovery in the morphology of the bioelectrical components of the SLAEP and even recovery from hearing loss[19], and although the prevalence in the general population has not been described, it is known to be more frequent in premature patients[19,20], so these patients have to be constantly monitored with SLAEP.

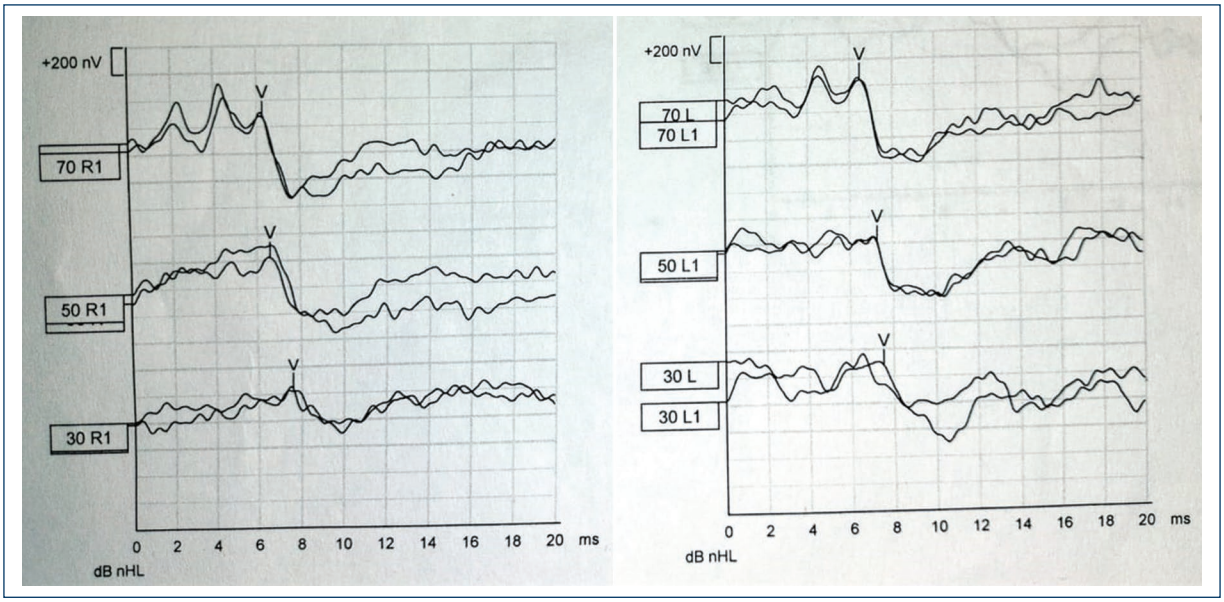


Figure 4. A 10-month record of SLAEP shows normal audiological and neurophysiological parameters.

Conclusion

All ANSD patients have in common absent or severely altered SLAEP, and present OAE, however the behavioral hearing threshold can only be determined by behavioral audiometric procedures, which is very difficult and unreliable to obtain in very young children. In ANSD behavioral thresholds can range from normal hearing to profound hearing loss. The only way to determine the need to adapt hearing aids in very young children is through the information the parents give us about speech development and auditory behavior, and frequent and close audiological surveillance as well.

Funding

The authors declare that they have not received funding.

Conflicts of interest

The authors declare no conflicts of interest.

Ethical considerations

Protection of humans and animals. The authors declare that no experiments involving humans or animals were conducted for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from the patient's parents, and received approval from the Ethics Committee. The SAGER guidelines were followed according to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that no generative artificial intelligence was used in the writing of this manuscript.

References

1. Starr A, Rance G. Auditory neuropathy. Hand Book Clin Neurol. 2015;129:495-508.
2. Van Straaten HL. Automated auditory brainstem response in neonatal hearing screening. Acta Paediatr Suppl. 1999;88:76-9.
3. Madden C, Rutter M, Hilbert L, Greinwald JH Jr., Choo DI. Clinical and audiological features in auditory neuropathy. Arch Otolaryngol Head Neck Surg. 2002;128:1026-30.
4. Baldwin SM, Gajewski BJ, Widen JE. An evaluation of the cross-check principle using visual reinforcement audiometry, otoacoustic emissions, and tympanometry. J Am Acad Audiol. 2010;21:187-96.
5. Sabo DL. The audiologic assessment of the young pediatric patient: the clinic. Trends Amplif. 1999;4:51-60.
6. Chinese Multi-center Research Collaborative Group on Clinical Diagnosis and Intervention of Auditory Neuropathy; Editorial Board of Chinese Journal of Otorhinolaryngology Head and Neck Surgery; Society of Otorhinolaryngology Head and Neck Surgery, Chinese Medical Association, China Division; International Association of Physicians in Audiology, Society of Audiology and Vestibular Medicine; China International Exchange and Promotive Association for Medical and Health Care. Chinese clinical practice guideline of auditory neuropathy (version 2022). Zhonghua Er Bi Yan Hou Tou Jing Wai Ke Za Zhi. 2022;57:241-62.
7. Maris M, Venstermans C, Boudewyns AN. Auditory neuropathy/dyssynchrony as a cause of failed neonatal hearing screening. Int J Pediatr Otorhinolaryngol. 2011;75:973-5.
8. Bansal S, Gupta A, Nagarkar A. Transient evoked otoacoustic emissions in hearing screening programs: protocol for developing countries. Int J Pediatr Otorhinolaryngol. 2008;72:1059-63.
9. Kai U. Transient auditory neuropathy in infants: how to conceptualize the recovery of auditory brain stem response in the context of newborn hearing screening? Semin Hear. 2011;32:123-28.

10. López-Díaz de León E, Silva-Rojas A, Ysunza A, Amavisca R, Rivera R. Auditory neuropathy in friedreich ataxia. A report of two cases. Int J Pediatr Otorhinolaryngol. 2003;67:641-8.
11. Rance G, McKay C, Grayden D. Perceptual characterization of children with auditory neuropathy. Ear Hear. 2004;25:34-46.
12. Bielecki I, Horbulewicz A, Wolan T. Prevalence and risk factors for auditory neuropathy spectrum disorder in a screened newborn population at risk for hearing loss. Int J Pediatr Otorhinolaryngol. 2012;76:1668-70.
13. Kumar UA, Jayaram MM. Prevalence and audiological characteristics in individuals with auditory neuropathy/auditory dys-synchrony. Int J Audiol. 2006;45:360-6.
14. Muthukumar R, Jaya V, Vignesh SS, Thenmozhi K. Prevalence and auditory characteristics of auditory neuropathy spectrum disorder in adult population with sensory neural hearing loss: a hospital based study in South India. Indian J Otolaryngol Head Neck Surg. 2023;75:1906-11.
15. Sahwan M, Abdelsamad Y, Alasfoor F, Alfayez F, Binkhamis G, Nichani J. Cochlear implantation in children with auditory neuropathy spectrum disorder: an updated systematic review. Eur Arch Otorhinolaryngol. 2024;281:1149-62.
16. Morlet T, O'Reilly R, Pritchett C, Venskytis E, Parkes W. A 15-year review of 260 children with auditory neuropathy spectrum disorder: II. Management and outcomes. Ear Hear. 2023;44: 979-89.
17. De Siati RD, Rosenzweig F, Gersdorff G, Gregoire A, Rombaux P, Deggouj N. Auditory neuropathy spectrum disorders: from diagnosis to treatment: literature review and case reports. J Clin Med. 2020; 9:1074.
18. Sharma A, Nash AA, Dorman M. Cortical development, plasticity and re-organization in children with cochlear implants. J Commun Disord. 2009;42:272-9.
19. Evans A, Baudoin M, Knight HE, Giles A. Case report: transient auditory neuropathy with resurgence of electrophysiologic waveforms observed between the neonatal period and age 3 years. J Am Acad Audiol. 2023;34:38-44.
20. Joint Commettee in Infant Hearing. Year 2019 position statement: principles and guidelines for early hearing detection and intervention programs. J Early Hear Detect Interv. 2019;4:1-44.