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İşitsel nöropati spektrum bozukluğu ve koklear implant

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ABSTRACT

Auditory neuropathy spectrum disorder is a pathology resulting from regional dysfunction extending from the auditory structures in the ear to the auditory brainstem. It is especially diagnosed by the presence of otoacoustic emission as well as the absence of auditory brainstem response or the presence of serious disorders in wave morphologies. Cochlear implant is one of the treatment methods for this disorder and is preferred by clinicians in order to contribute to speech perception and language development. Here, some cochlear implant candidacy criteria come into play and the implantation option is applied if the necessary conditions are met for the auditory neuropathy spectrum disorder patient population. In this study, the effects of cochlear implant on auditory neuropathy spectrum disorder were compiled by including the researches of the last 10 years. As a result of the studies, it was determined that the cochlear implant improved postoperative speech perception and contributed positively to language development in the auditory neuropathy spectrum disorder population without the presence of any comorbidity. However, more studies with high evidence are needed for the effect of cochlear implant on auditory neuropathy spectrum disorder in the presence of comorbidity.

Keywords: auditory neuropathy spectrum disorder, cochlear implant, speech perception, hearing aid

ÖZ

İşitsel nöropati spektrum bozukluğu ve koklear implant

İşitsel nöropati spektrum bozukluğu, iç kulakta bulunan işitsel yapılardan işitsel beyin sapına kadar uzanan bölgedeki disfonksiyondan kaynaklanan bir patolojidir. Özellikle otoakustik emisyon varlığı ve bunun yanında işitsel beyin sapı cevabının alınamaması veya dalga morfolojilerindeki ciddi bozuklukların varlığı ile tanılanmaktadır. Bu bozuklukta, işitme cihazı ve koklear implant uygulaması gibi bazı amplifkasyon/tedavi yaklaşımları uygulanmaktadır. Koklear implant, konuşma algısı ile dil gelişimine katkıda bulunması adına klinisyenlerce tercih edilmektedir. Burada bazı koklear implant adaylık kriterleri devreye girmekte ve işitsel nöropati spektrum bozukluğu hasta popülasyonu için gerekli şartlar sağlanması halinde implantasyon seçeneği uygulanmaktadır. Bu derlemede koklear implantın işitsel nöropati spektrum bozukluğundaki etkileri son 10 yılın araştırmaları dâhil edilerek derlenmiştir. Çalışmaların ortak sonucunda, koklear implantın herhangi bir komorbidite varlığı olmadan işitsel nöropati spektrum bozukluğu popülasyonuna postoperatif konuşma algısını geliştirdiği ve dil gelişimine olumlu yönde katkı sunduğu yayınlara bldirilmiştr. Ancak koklear implantın komorbidite varlığında işitsel nöropati spektrum bozukluğuna etkisi adına kanıt değeri yüksek daha fazla çalışmaya ihtiyaç vardır.

Anahtar kelimeler: işitsel nöropati spektrum bozukluğu, koklear implant, konuşma algısı, işitme cihazı

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INTRODUCTION

Auditory neuropathy spectrum disorder (ANSD) is a disease in which the transmission of auditory signals from the inner ear to the auditory nerve and auditory brain stem is impaired (Manchaiah, Zhao, Danesh, & Duprey, 2011). Diagnosis can be made by ABR and OAE features, which are electrophysiological test methods. The presence of normal OAE or cochlear microphonic responses, as well as the absence of a decreased Wave 5 or Wave 5 in ABR responses, leads clinicians to the diagnosis (Siati, Rosenzweig, Gersdorff, Gregoire, Rombaux, & Deggouj, 2020) (Roche, Huang, Castillo, Bassim, Adunka, & Buchman, 2010). Symptoms of the disease include fluctuating hearing sensitivity, difficulty in hearing in noisy environments, and poor speech perception performances (Teagle, Roush, Woodard, Hatch, Zdanski, Buss, & Buchman, 2010).

The prevalence of the disorder is estimated to vary between 0.5% and 15% in all patients evaluated with hearing loss (Mittal, 2012). In a study, the incidence of ANSD was found to be 0.09 in 1000 infants (Boudewyns, Declau, van den Ende, Hofkens, Dirckx, Van de Heyning, 2016). Prematurity and jaundice (hyperbilirubinemia) were found among the most common etiological factors observed in a study (Rajput et al., 2019).

The pathophysiological mechanism of auditory neuropathy is examined in 4 parts. These are:

1. Presynaptic disorders affecting inner hair cells and synapses,

2. Postsynaptic disorders affecting unmyelinated auditory nerve dendrites,

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3. Postsynaptic disorders affecting auditory ganglion cells and their myelinated axons and dendrites,

4. Disturbances in the central neural pathways affecting the auditory brain stem (Rance & Starr, 2015).

Among the evaluation methods, ANSD can be diagnosed using objective test methods such as otoacoustic emission (OAE) or cochlear microphonic measurements (CM), auditory brainstem response (ABR). Especially the absence of ABR responses with the presence of OAE may indicate that there is a pathology after the superior oliveric complex (SOC) level (Kaga, 2016; Norrixa & Velenovskya, 2014). Because auditory evoked cortical responses require less neural synchronization than auditory brainstem (ABR) responses, these potentials are achieved in some of the cases (Belgin & Şahlı, 2017; Rance, 2005). Other audiological findings are hearing loss in different degrees and configurations, mostly bilateral, speech discrimination scores being quite low compared to audiograms, tympanometric measurement result being within normal limits because the location of the pathology is in the auditory path beyond the middle ear, and acoustic reflexes are generally not obtained because spiral ganglion cells and/or auditory nerve fibers are affected (Evren, Demirbilek, & Küfeciler, 2021; Raveh, Buller, Badrana, & Attias, 2007; Sininger & Oba, 2001).

There are three current rehabilitation modality for children with ANSD: hearing aids (HA), frequency modulated technology and cochlear implants (CI) (Walker, McCreery, Spratford, & Roush, 2016).

Hearing loss affects the communication and speaking skills of the individual negatively, regardless of age (Özcan, Batuk, Kaya, & Sennaroğlu, 2021). Cochlear implantation has become the standard treatment for adults and children with severe to profound hearing loss (Shearer & Hansen, 2019). Cochlear implant is a system that electrically stimulates the auditory nerve, used in individuals with severe to profound sensorineural hearing loss (Sennaroğlu, Batuk, & Kaya, 2019). Cochlear implantationin our country for the first time, it was done by Dr. Bekir Altay in 1987 at Anadolu University Medicosocial Hospital (Altay & Konrot, 2006).

The aim of this study is to reveal the effect of cochlear implant on postoperative speech perception and language development of auditory neuropathy spectrum disorder cases by compiling studies conducted in the last 10 years.

ANSD and CI

CI Candidate Criteria

Selecting patients suitable for cochlear implant surgery among children and adults who cannot make sufficient progress in speech understanding, speech and language development after receiving adequate auditory rehabilitation/habilitation and/or acoustic amplification; Age at implantation, radiological evaluations, genetic tests to determine whether otoferlin gene mutation is present, pure tone audiometry, hearing aid experience and ECochG evaluations are the parameters that should be evaluated before cochlear implantation (CI) candidacy (Raza, Aryal, Prabhu, 2023; Sahwan, Abdelsamad, Alasfoor, Alfayez, Binkhamis, & Nichani, 2023).

All children with ANSD undergo a conventional hearing aid trial for at least 6 months without any significant benefit in speech perception and language development to be considered CI (Kontorinis, Lloyd, Henderson, Jayewardene-Aston, Milward, Bruce, & Freeman, 2014). The hearing thresholds of children with ANSD are not low and are even much better than children in the CI nomination criteria, but the language development and speech comprehension levels of children with ANSD are significantly behind their peers (Harrison, Gordon, Papsin, Negandhi, & James, 2015). Selection of patients suitable for cochlear implant surgery among children and adults diagnosed with ANSD who are able to respond to pure-tone audiometry testing; According to pure tone audiometry and speech test results with hearing aid, it is determined whether it is an implant candidate or not (Musiek, Baran, Shinn, & Jones, 2011).

While it was previously accepted that only children with bilateral profound hearing loss (>90 dB) who received little or no benefit from hearing aids should be considered for implantation, the criteria were later expanded to include individuals with severe hearing loss and residual hearing (Alkaya. 2022). Implant application recommendations:

1. For children with deep (>90 dB HL) bilateral SNHL in children under 2 years of age, or with severe to profound bilateral SNHL in children aged 2 to 17 years, in both cases, little or no benefit despite properly fitted hearing aids is sufficient to recommend implant application.

2. Individuals with normal to moderate pure tone thresholds should consider implants if their speech perception and auditory processing are significantly lower and there is limited benefit from conventional amplification (Varadarajan, Sydlowski, Li, Anne, & Adunka, 2021).

In our country, the social security institution frequently updates the payment scope of cochlear implant application. The latest SUT (Health Practice Communiqué) criteria for those diagnosed with auditory neuropathy published in the Official Newspaper on 16 March 2023, the latest update, are as follows: In cases diagnosed with auditory neuropathy; If it is documented with an audiological evaluation and a health board report that he has not benefited from hearing rehabilitation and education for at least 6 months, the cost is covered by the institution.

Researches on Cochlear Implant and Auditory Neuropathy Spectrum Disorder

A retrospective study by Budenz et al. included 26 CI users with ANSD and compared them in terms of speech perception before

and after surgery. Seventeen CI children with isolated ANSD were compared with a similar group of children with hearing loss, and 9 children with a diagnosis of ANSD associated with cognitive or developmental disorders were compared with children with isolated ANSD. As a result of the study, 17 children with isolated ANSD performed comparable to their peers, while children with ANSD associated with cognitive or developmental disorders showed worse CI performance compared to children with isolated ANSD (Budenz, Telian, Arnedt, Starr, Arts, El-Kashlan, & Zwolan, 2013).

In another study conducted in Brazil, 15 CI users with an average age of 10 and diagnosed with ANSD were included and their speech perceptions were evaluated with the noise hearing test (HINT). As a result of the study, it was concluded that the use of CI improves auditory skills in individuals with ANSD and has the ability to improve neural synchronization for auditory rehabilitation in this population (Fernandes & Bevilacqua, 2013).

Ching et al. evaluated the performance levels (speech, language, and psychosocial development) of 47 children with ANSD. Twenty-seven children were HA and 19 children were cochlear implant users. As a result of the study, it was revealed that there was no significant difference in performance levels or variability for children between children with and without ANSD (Ching, Day, Dillon, Gardner-Berry, Hou, Seeto, & Zhang, 2013).

Dean et al. evaluated the speech perceptions of 27 children with ANSD who were unilateral and bilateral CI users without any additional inner ear anomalies. As a result of the study, it was revealed that children with bilateral CI users had better speech perception. Poor performers were found to have a later implantation age and lower socio-economic status. The study revealed that children with ANSD will be successful in education and habilitation if they are implanted at a young age if they do not benefit from conventional devices (Dean, Felder, & Kim, 2013).

In a study by Jeong and Kim, the postoperative speech perception abilities of 15 CI users diagnosed with ANSD were evaluated using radiological imaging and electrophysiological measurements. As a result of the study, postoperative speech perception abilities were found to be excellent in patients with normal bone cochlear nerve canal (BCNC) and normal cochlear nerve findings. In addition, poor speech perception was observed in patients with a narrow or obliterated BCNC and cochlear nerve insufficiency (Jeong & Kim, 2013).

A retrospective study by Pelosi et al. on the subject included 26 children with ANSD. Cochlear implantation was applied to 16 patients and binaural HA was applied to 10 patients, and their speech perception performances were evaluated after the application. As a result of the study, ANSD patients with CI users were able to achieve overall speech perception results comparable to hearing aid wearers only (Pelosi, Wanna, Hayes, Sunderhaus, Haynes, Bennett, & Rivas, 2013).

In a related review examining 27 studies, the speech recognition performance of CI users with ANSD and hearing aid users was found to be equivalent. However, at the end of the study, it was stated that stronger evidence and research are needed due to the weakness of the evidence and methodological concerns (Humphriss, Hall, Maddocks, Macleod, Sawaya, & Midgley, 2013).

In a retrospective study examining the medical reports of patients with ANSD who were treated with cochlear implants between 2000 and 2012, 27 children (17 boys, 10 girls) were diagnosed with ANSD as a result of ABR and OAE tests. After the children used conventional HA, unilateral CI was applied to 18 children and bilateral CI was applied to 9 children. Comorbidity was detected in 5 children. As a result of the study, if the presence of comorbidity is excluded, it was seen that the use of CI for auditory rehabilitation in children with isolated ANSD (only individuals with ANSD without comorbidity) can be beneficial (Kontorinis et al., 2014).

In a study conducted by Sarankumar et al., investigating the effect of CI on ANSD, auditory and speech scores of 10 patients (8 men and 2 women) with ANSD were compared after 12 months of habilitation. Study outcome was evaluated by auditory evoked cortical potentials, and P1 latency in children with ANSD was well correlated with auditory and speech scores. As a result of the study, significant benefits were seen in children with ANSD who underwent CI one year after habilitation (Sarankumar, Arumugam, Goyal, Chauhan, Kumari, & Kameswaran, 2018).

In the systematic review of Chaudhry et al. on the subject, 14 studies were included. In the common denominator of the studies, it was determined that the majority of the patients benefited from CI and that even in patients with syndromes in addition to ANSD, improvement in hearing thresholds and speech perception was observed after CI (Chaudhry, Chaudhry, Muzaffar, Monksfield, & Bance, 2020).

In a mini-systematic review by Myers and Nicholson, articles evaluating children with ANSD who underwent CI before the age of 3 were reviewed. As a result of the study, it was stated that CI application is a suitable intervention method for children with ANSD and similar outcome performances were observed for children with other sensorineural hearing loss (Myers & Nicholson, 2021).

The study of Lin et al., which aimed to investigate the results of CI in pediatric patients with ANSD of different etiologies, included 36 children with ANSD who underwent CI between 2001 and 2021. The etiological causes of the children were analyzed using next-generation sequencing-based genetic studies, high-resolution computed tomography and magnetic resonance imaging (MRI). As a result of the study, insufficient CI results were obtained in patients with ANSD with cochlear nerve insufficiency (Lin et al., 2022).

Information about these studies and a summary of the results are shown more comprehensively in table 1 and Table 2.

Table 1. Summary of informations on included studies

Study	N	Age at diagnosis	Presence of comorbidity	Previous intervention	Preoperative hearing loss severity
Budenz et al., 2013	26	*Iso-Gr, Co-Gr* 14–85, 13–85 months	Cognitive or developmental disorders (n=9)	Hearing aid and rehabilitation	Profound (26)
Ching et al., 2013	47	2.2 months	NR	Hearing aid	Mild (6), Moderate (18), Severe (6), Profound (17)
Dean et al., 2013	27	2.5–9.7 years	History of ear infections and ear anomalies (n=7) Developmental disorders (autism) (n=1)	Hearing aid	NR
Jeong & Kim, 2013	15	NR	Obliterated Bony of Cochlear Nerve (n=3)	NR	Profound (15)
Pelosi et al., 2013	26	4–23 months	Motor/cognitive developmental delay (n=8) Developmental delay (n=5)	Hearing aid	Mild-profound (2) NR
Kontorinis et al., 2014	27	NR	Cognitive disorders (n=4) Co-morbidities (n=1)	Hearing aid	Profound
Sarankumar et al., 2018	10	NR	NR	Hearing aid	Profound

Co-Gr: ANSD patients group with comorbidities; Iso-Gr: isolate ANSD patients group; NR: not recorded.

Table 2. Studies examining cochlear implantation (CI)

Study	N	Age at Intervention	Outcome Measured	Pre-CI	Post-CI
		Iso-Group, Co-Group		Iso-Group, Co-Group	Iso-Group, Co-Group
Budenz et al., 2013	26	14–85, 17–90 months	AV Oral TC ASL IT-MAIS score (%)	35.3%, 29.4% 47.1%, 58.8% 11.8%, 11.8% 5.9%, 0 NR	35.3%, 0 47.1%, 11.1% 11.8%, 88.9% 5.9%, 0 38–88%, 15–45%
					SNHL, ANSD
Ching et al., 2013	33	6.6 months	PLS-AC PLS-EC PPVT DEAP_C DEAP_V PEACH	NR	24.6, 35.9 19.4, 17.8 16.7, 16.4 2.6, 2.5 2.69, 2.1 21.0, 23.2
					After 6–12–24 months
Dean et al., 2013	27	3.4–14.8 years	Speech Perception Scores (%)	67%	86%, 79%, 87%
Jeong & Kim, 2013	15	1.1–7.5 years	MWT for Phoneme MWT for Word IT-MAIS (%)	*Good gr., Poor gr.* 0–11%, 0–32% 0 2.5–10%, N	*Good gr., Poor gr.* 63–100%, 0–49% 34–100%, 0–30% 100%, NR
		CI Group, HA Group		CI Group, HA Group	CI Group, HA Group
			IT-MAIS scores (%) SRT (dB) NU-CHIPS	13.5–65, 3–72% 30–100, 30–70 dB	60–100, 70–88%
Pelosi et al., 2013	24	1–10.5, 1–6 years	Score (%) HINT-C Score (%) NU-6 (%) PBK (%)		63–98, 96–98% NR, 68% NR, 100%
Kontorinis et al., 2014	27	19–68 months	CAP MSLDS	2.5 (range 0–5) 2.5 (range 0–6)	5.8 (range 2–9) 7.7 (range 3–10)
				ANSD Gr., SNHL Gr.	ANSD Gr., SNHL Gr.
Sarankumar et al., 2018	10	1.5–6 years	CAP SIR MAIS MUSS SPIN Scores 0 dB SNR (%) SPIN Scores +10 dB SNR (%)	0–2, NR 1, NR NR NR NR NR	4–6, 4–6 1–5, 2–5 30–39, 32–40 12–29, 24–33 55–70%, NR 65–90%, NR

ANSD: auditory neuropathy spectrum disorder; ASL: American Sign Language; AV: auditory verbal communication; CAP: categories of auditory performance; CI: cochlear implantation; Co-Gr: ANSD patients group with comorbidities; dB: decibel; DEAP-C: consonant correct score; DEAP-V: vowel correct score; HA: hearing aid; HINT-C: Hearing in Noise Test sentences for children; Gr.: group; Iso-Gr: isolate ANSD patients group; IT-MAIS: Infant-Toddler Meaningful Auditory Integration Scale; MAIS: meaningful auditory integration scale; MSLDS: Manchester spoken language development scale; MUSS: meaningful use of speech score; MWT: monosyllabic word test; NR: not recorded; NU-CHIPS: Northwestern University–Children's Perception of Speech; NU-6:Northwestern University Auditory Test No. 6; PBK: Phonetically Balanced Kindergarten; PEACH: Parents evaluation of aural/oral performance of children; PLS-AC: auditory comprehension and score; PLS-EC: expressive communication score; PPVT: receptive vocabulary score; SIR: speech intelligibility rate; SNHL: sensorineural hearing loss; SNR: signal noise ratio; SPIN: speech perception in noise; SRT: speech reception threshold; TC: total communication.

CONCLUSION

Auditory neuropathy spectrum disorder is a condition that develops with a wide variety of pathophysiological mechanisms. The presence of additional behavioral and developmental comorbidities arising from neuropathy in patient groups with ANSD, as well as factors such as anatomical anomalies (nerve hypoplasia, etc.) accompanying ANSD, cause outcome measurement studies to be inconclusive enough to provide prognostic information. Therefore, in the studies compiled in this study, the presence of comorbidity in the ANSD population does not make a significant difference in performance after CI and does not offer significant improvement to auditory rehabilitation. In addition, as a result of the studies, it was stated that the results were equivalent to the other groups compared in the speech perception performances after CI, especially in the isolated ANSD patient group. This result shows the clinicians that the implantation option is appropriate in terms of speech perception and language development in ANSD patients without comorbidity (isolated ANSD), and additionally, it has been reported that more studies with a high level of evidence are needed for individuals with ANSD in the presence of comorbidity.

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