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Pediatric sudden sensorineural hearing loss: etiology, prognostic factors and treatment

Çocuklarda ani sensörinöral işitme kaybı: etiyoloji, prognostik faktörler ve tedavi sonuçları

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ABSTRACT

Aim: Sudden sensorineural hearing loss (SSNHL) may have a negative impact on the language and psychological development of children, especially if it is not diagnosed early and treated promptly. This study were aimed to determine and compare the etiological factors, treatment outcomes and prognostic factors in the pediatric patients who were followed up with the diagnosis of SSNHL.

Materials and Methods: The files of 28 children were analyzed retrospectively. In pure tone audiometry, the average of pure tone thresholds of 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz were accepted as pure tone averages (PTAV). Patients' recovery status was determined according to Siegel criteria. Audiometric curve types were evaluated as ascending, descending, and flat.

Results: The ages of patients with SSNHL was mean ± SD 14.89 ± 3.24 (min-max: 7 and 18). The pre-treatment PTAV was mean ± SD 55.27 ± 12.39 dB HL (min-max: 38.5 and 85.25 dB HL) and the PTAV after treatment was mean ± SD 23.13 ± 18.22 dB HL (min-max: 5 and 72.5 dB HL). Audiometric curve types were detected as descending curve (n: 11, 39.2%), ascending curve (n: 5, 17.9%), flat curve (n: 12, 44.9%). Eighteen (64.3%) patients had complete recovery, 8 (28.6%) patients had partial recovery, and 2 (7.1%) patients had no recovery after the treatment.

Conclusion: The response to treatment was found to be high. Descending audiometric curve type was found as a positive prognostic factor. Although the presence of tinnitus was a better prognostic factor than the presence of vertigo.

Keywords: Hearing loss, sudden hearing loss, prognosis, child, adolescent.

ÖZ

Amaç*: Geç teşhis edilen ve tedavi ile düzelmeyen ani sensörinöral işitme kaybı (ASNİK) çocukların dil ve psikolojik gelişimi üzerinde olumsuz bir etkiye sahip olabilir, bu nedenle tanı konur konulmaz tedaviye başlanmalıdır. Bu çalışmanın amacı ASNİK ile takip ve tedavi edilen çocuk hastalarda etiyoloji, prognostik faktörlerin belirlenmesi ve tedavi sonuçlarının karşılaştırılmasıdır.*

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***Gereç ve Yöntem**: ASNİK tanısı ile tedavi ve takip edilen 28 hasta çocuğun dosyaları retrospektif olarak incelendi. Pür ton odyometrisinde (PTO), 500 Hz, 1000 Hz, 2000 Hz ve 4000 Hz saf ton eşiklerinin ortalaması saf ses ortalaması (SSO) olarak kabul edildi. Hastaların iyileşme durumları Siegel kriterlerine göre belirlendi. Odyometrik eğri türleri artan, azalan ve düz tip olarak gruplara ayrıldı.*

***Bulgular**: ASNİK'li hastaların yaşları 7 ile 18 arasında değişmekteydi (ortalama 14,89 ± 3,24). Tedavi öncesi SSO 55,27 ± 12,39 dB (min-maks: 38,5 ila 85,25 dB), tedavi sonrası SSO 23,13 ± 18,22 dB (min-maks: 5 ila 72,5 dB) idi. Odyometrik eğri türleri: azalan grup (n: 11, %39,2), yükselen grup (n: 5, %17,9), düz grup (n: 12, %44,9) şeklinde idi. Tedavi sonrası 18 (%64,3) hastada tam iyileşme, 8 (%28,6) hastada kısmi iyileşme saptanırken 2 (%7,1) hastada iyileşme olmadı.*

***Sonuç**: Çalışmamızda sistemik steroid ile tedaviye yanıt oranı yüksek saptandı. Kulak çınlaması varlığı ve tedavi öncesi inen tip odyometrik eğri iyi prognoz faktörleri olarak saptandı.*

***Anahtar Sözcükler:** İşitme kaybı, ani işitme kaybı, prognoz, çocuk, adolesan.*

INTRODUCTION

Sudden sensorineural hearing loss (SSNHL) is defined as a sensorineural hearing loss of 30 dB or greater over at least three consecutive frequencies occurring within three days and is accepted as an otorhinolaryngological emergency (1). The incidence of SSNHL is reported to be 5-20/100,000, but the actual incidence rate may be higher as patients do not apply to the hospital because of the high rate of spontaneous recovery (1). Although it can be seen in all age groups, it is rare in childhood and the elderly (1-3). Alexander and Harris (4) reported the incidence as 11/100,000 in children under 18 years of age and 27/100,000 in patients over 65 years of age. Most of the cases are unilateral; less than 2% of patients have bilateral hearing loss (3-6). A variety of diseases and factors have been proposed that may lead to SSNHL; while some of them can be revealed with anamnesis, physical examination and tests, specific etiologies can only be found in 10% of cases. When a precise etiology is not found despite proper investigation, it is called idiopathic SSNHL and 90% of the patients are idiopathic (7, 8).

Childhood is the period for the development of speech and language; therefore, without appropriate treatment, SSNHL can cause serious problems such as permanent hearing loss and can deeply impact the cognitive development of children, leading to psychiatric sequelae, which increase the burden on families and society.

The aim of this study was to determine and compare the etiology, laboratory results, prognostic factors and treatment outcomes in children with SSNHL, by evaluating the results of one of the largest pediatric SSNHL series in our country.

MATERIALS and METHODS

Study design and patients

Ethics committee approval was obtained for the study (Decision no: 01/05.09.2018). This study was carried out in accordance with the principles of the Declaration of Helsinki. The files of 28 children, between the ages of 7-18 years who were admitted to our hospital with the diagnosis of SSNHL and followed up at least three months between January 2008 and December 2017 were retrospectively analyzed.

While patients with pre- and post-treatment third month pure tone audiometries were included to the study, patients with middle ear or retrocochlear pathology, autoimmune inner ear disease, Meniere's disease, syndromic or genetic hearing loss, history of ear surgery were excluded.

Audiometric assessment

In pure tone audiometry, the average pure tone thresholds of 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz were accepted as pure tone averages (PTAV). The severity of hearing loss was based on the PTA as follows;

1. 25-40 dB HL mild,
2. 41-55 dB HL mild-moderate,
3. 56-70 dB HL medium,
4. 71-90 dB HL severe.

Patients' recovery status was determined according to Siegel's criteria (9).

Three types of SSNHL was defined according to audiogram shape: ascending, descending and flat.

Treatment protocols

All patients were treated with systemic steroids. Oral methylprednisolone was administered at a

dose of 1 mg/kg and tapered in 14 days. Patients who did not improve or had a PTA greater than 60 dB HL at the fifth-day control audiogram, were injected three doses (1 dose=0.5 ml=2 mg) of intratympanic dexamethasone solution (Dekort, DEVA Corporation, Istanbul, Turkiye) every other day if they tolerated the injection. Patients with a history of upper respiratory tract infection were tested for CMV, EBV, Mumps, Herpes viruses (HSV, VZV) and Influenza. An antiviral treatment was initiated in patients with positive viral serology.

Statistical Analysis

Descriptive statistics were calculated for all variables, an association between the groups were evaluated using the Chi-square test. The Independent Samples t-test was used to compare the means of two independent groups and the nonparametric Mann-Whitney U test was applied to investigate continuous variable prognostic factors. All statistical analyses were performed using SPSS version 22.0 (IBM SPSS Statistics, Chicago, IL, USA). A p-value less than 0.05 was considered as statistically significant.

RESULTS

Patient characteristics

Twenty-eight children were treated with the diagnosis of SSNHL during the study period. Five patients had a possible etiologic factor; one had a history of acoustic trauma, one was treated with streptomycin for brucellosis, one had Type 1 Diabetes Mellitus (DM), two patients with a history of upper respiratory infection had positive viral serology. The remaining 23 patients (82.1%) were diagnosed as idiopathic SSNHL. The ages of patients with SSNHL was mean ± SD 14.8 ± 3.2 years (min-max: 7 and 18 years) (Table-1). The follow-up time was mean ± SD 15.3 ± 6.0 months (min-max: 3 and 26 months). The time passed from hearing loss and initiation of the treatment was mean ± SD 2.7 ± 1.7 days (min-max: 1 and 7 days) (Table-1).

Twelve of the patients were boys and 16 were girls. While the hearing loss was on the right side in 53.6% (n = 15) of the children, it was on the left side in 39.3% (n = 11). Hearing loss was bilateral in one of the patients (3.6%) (Table-2). Five patients (17.9%) had a history of upper respiratory tract infection. Eight patients had tinnitus and three patients had vertigo accompanying hearing loss (Table-2). The degrees of sensorineural hearing loss were; 10.7% (n=3) mild, 57.1% (n=16) mild-moderate, 28.6% (n=8) moderate, 3.6% (n=1) severe. Twenty-five patients received oral steroids, three patients received oral steroids + intratympanic steroids and three patients with positive viral serology received oral steroids + antiviral treatment. There was no improvement in two of 3 patients with vertigo and partial recovery was observed in one patient (Table-2). Of the eight patients with tinnitus, six had complete recovery, and two had partial recovery. Only one patient was treated with the diagnosis of bilateral SSNHL. The patient was a 17-year-old boy; pre-treatment PTA values were 44.75 dB HL for the right ear and 42.50 dB HL for the left ear. On the third month of the treatment, PTA values were 14 dB HL for the right ear and 13.50 dB HL for the left ear.

All patients underwent Magnetic Resonance Imaging (MRI) and internal acoustic canal dilatation was found in one patient. No other specific findings were observed in MRI's. Temporal bone computed tomography was also performed in 10 patients, and no pathological lesions were observed either.

Table-1. Descriptive analysis.

	Mean ± SD (Minimum-Maximum)
Age (years)	14.8 ± 3.2 (7-18)
Initial treatment time (day)	2.7 ± 1.7 (1-7)
Follow-up (month)	15.3 ± 6.0 (3-26)
PTA-pre (dB HL)	55.2 ± 12.4 (38.5-85.2)
PTA-post (dB HL)	23.1 ± 18.2 (5-72.5)

Standard deviation (SD), decibel (dB), hearing level (HL), pure tone averages (PTAV),

Table-2. Clinical characteristics and prognostic factors.

	Complete improvement (n=18)	Partial + No improvement (n=10)	P values
Age (years) Mean ± sd	15.67 ± 2.33	13.50 ± 4.25	0.208
Gender (Female: F, Male: M, n, %)	9 (50%), 9 (50%)	7 (70%), 3 (30%)	0.434
Side (R: Right, L: Left, B: Bilateral, n, %	R= 9 (50%) L= 8 (44.4%) B=1 (5.6%)	R=6 (60%) L= 4 (40%) B=0 (0.0%)	0.540
Vertigo (n, %)	2 (11.1%)	1 (10%)	0.130
Tinnitus (n, %)	4 (22.2%)	4 (40%)	0.400
Ear fullness (n, %)	1 (5.6%)	2 (20%)	0.284
Initial treatment time (mean ± sd day)	2.78 ± 1.83	2.60 ± 1.43	0.960
Degree of hearing loss (mean ± sd)	1.17 ± 0.62	1.40 ± 0.84	0.480
PTAV pre (mean ± sd dB HL)	52.33 ± 11.07	60.58 ± 13.46	0.160

Pure tone averages (PTAV), standard deviation (sd), decibel (dB), hearing level (HL),

Treatment outcomes

The pre-treatment PTAV was mean ± SD 55.2 ± 12.4 dB HL (38.5 and 85.2 dB HL), post-treatment PTAV was mean ± SD 23.1 ± 18.2 dB HL (min-max: 5 to 72.5 dB HL) (Table-1).

Eleven patients had a descending curve (39.2%), five patients had an ascending curve (17.9%) and 12 patients had flat curves (44.9%). Audiological outcomes revealed that descending audiogram type was positively associated with hearing recovery (p=0.017).

According to the results of post-treatment PTA, which was performed at least three months after the treatment, 18 (64.3%) patients had complete recovery, eight (28.6%) patients had partial recovery, and two (7.1%) patients had no improvement (Table-2).

Prognostic factors

We divided the patients into two groups; complete recovery and partial recovery + no improvement group according to Siegel's criteria. There was no significant difference between the groups in terms of age, gender and side (p>0.05) (Table-2). In patients with tinnitus, response to treatment was better than patients with vertigo (p = 0.022, p=0,086, respectively). There was no statistical difference between treatment initiation time and treatment response rate (p> 0.05) (Table-2). No improvement was observed in two of 3 patients who received intratympanic steroid injections.

Serologic tests were done in five patients with a history of upper respiratory tract infection, Cytomegalovirus (CMV) and Mumps IgM were positive in two patients. For these two patients, famciclovir was added to the systemic steroid treatment for one week. On the 10th day of treatment, the control audiogram showed complete recovery in both patients.

Of the 18 patients who showed complete recovery, four had tinnitus and two had vertigo. No accompanying symptoms were found in the rest of the patients. Of the eight patients who showed partial recovery, four had tinnitus and one had vertigo (Table-2). No significant difference was found between the complete recovery and partial and no improvement groups in terms of the presence of tinnitus and vertigo (p=0.400 and p=0,130, respectively).

DISCUSSION

The etiology and treatment of SSNHL have been discussed for many years and numerous different treatment protocols have been proposed in the literature (7, 8, 10, 11, 12). Although some studies reported that the recovery rate without treatment is over 50% in adult patients with sudden hearing loss (10, 11), it is clear that the treatment should be initiated as soon as the diagnosis is made in pediatric patients with SSNHL to avoid undesirable consequences.

Steroids, antioxidants, vasodilators and antivirals are commonly used for treatment, but the only agent whose efficacy is recognized worldwide, and proven to be effective, are steroids (11, 12). Although steroids are usually used systemically, intratympanic steroid administration which aims to avoid side effects of systemic steroid use by directing them to the target organ has been common in recent years (13, 14). The success rates of the intratympanic steroid administration (in combination with systemic steroid or as salvage therapy) varies widely so far (13, 14). Lim et al. (15) and Övet et al. (16) reported no significant difference between hearing results of the groups that received systemic oral steroid or combined therapy (steroid + intratympanic dexomethasone). We used intratympanic dexomethasone only in cases with inadequate or no response to oral steroids, but no improvement was observed in the hearing of two of 3 patients. However, the low number of patients prevents us from making a final judgment on the effectiveness of intratympanic steroid injection in pediatric patients.

Infectious, immunological, inflammatory causes and microcirculation insufficiency are the most important proposed etiological factors of SSNHL (14, 17, 18). Hereditary prothrombotic conditions that cause insufficiency of ear microcirculation by increasing blood viscosity such as diabetes and hypertension may lead to SSNHL (14, 17, 18). We also had a patient with Type 1 Diabetes Mellitus (DM), who had been receiving treatment for six years, and he showed partial recovery.

Pitaro et al. (19), in their study that included 19 patients with pediatric SSNHL, detected CMV in six patients and Epstein - Barr virus in one patient. They used antivirals in addition to oral steroids in serologically positive patients and achieved a complete recovery in one patient and partial recovery in four patients after the treatment. We only initiated, antiviral treatment in two patients with positive viral serology with the recommendation of the infectious diseases physician. Complete recovery was achieved in both patients after treatment. Antivirals seem to be effective in addition to steroids if serology is positive in children.

Several studies have been performed on pediatric SSNHL in our country. Kızılay and Koca (3) reported complete recovery in only three (21.4%) children. No recovery was seen in 11 (79.6%) patients, which were all under 15 age. Their low recovery rate, which is lower than our results as well as the other studies reported in the literature, is probably due to the very high rate of profound hearing loss in their study group. Ten (71.4%) of their patients had total hearing loss, while two had mild to severe hearing loss.

Övet G. et al (16), divided 49 pediatric patients into two groups in the study comparing systemic steroid treatment alone with combined steroid treatment (systemic steroid + intratympanic steroid injection). 75.5% (37 patients) of their patients showed complete + partial recovery, with no significant difference between treatment modalities.

In their study investigating the prognostic effects of neutrophil-lymphocyte ratio and platelet-lymphocyte ratios on pediatric SSNHL, Bulğurcu S. et al. (20) investigated 21 patients with idiopathic SSNHL under the age of 19 years. They reported complete + partial recovery in 12 (57.1%) patients, and neutrophil-lymphocyte ratio was a positive prognostic factor, similar to the results performed among adult patients with SSNHL. No significant difference was found for platelet to lymphocyte ratio in terms of recovery rates.

Overall (complete + partial + slight) recovery rates in pediatric SSNHL were reported between 21.4% and 75.5% in the literature (3, 16, 17, 20, 21). The overall recovery rate was 92.6% in our study population, which may be related to the low number of patients, absence of accompanying diseases, the high compliance rate of the patients to the treatment, as all the patients had completed the treatment and follow-up protocol, or to the early initiation of the treatment since all patients received treatment in the first seven days of the hearing loss.

Previous studies reported higher recovery rates in patients that received early initial treatment and ascending curve type of audiogram (21, 22). In contrast, higher recovery rates in pediatric

patients with descending curve type audiograms have also been reported (17). We did not find a significant impact of treatment initiation time on recovery rates but this is probably due to the initiation of the treatment in the early period of hearing loss in all of our patients. We also found that descending curve type is associated with better healing rates.

Skarzynski et al. (22) stated in their review that, while unilateral hearing loss, tinnitus, mild/moderate hearing loss, early beginning to treatment, ascending type pure tone audiometry configuration were positive prognostic factors, vertigo, bilateral hearing loss, delayed treatment, severe to profound hearing loss, descending type configuration of the audiogram were negative prognostic factors. Also, they reported that the most common etiological factor was idiopathic; viral infections, acoustic trauma, use of ototoxic drugs, rarely large vestibular aqueduct and congenital anomalies were also among etiologic factors. Another systematic review on the possible etiologies of SSNHL in adult patients showed that the etiology was idiopathic in 71% of cases (23). Similarly, most of the cases were idiopathic in our study. Also, even tinnitus was a better prognostic factor than vertigo in the current study, both symptoms did not show any prognostic impact on recovery rates. This contradiction may be attributed to low number of patients in our study, but it must be kept in mind that tinnitus and vertigo are both subjective symptoms that are difficult to evaluate, especially in the pediatric population. One of the limitations of our study is the absence of patients receiving hyperbaric oxygen therapy.

CONCLUSION

Sudden sensorineural hearing loss in children is a rare health condition and the current study is one of the largest series reported from our country. We found that the response to treatment with systemic steroid therapy was high in children and It can be said that those with tinnitus have a higher recovery rate than those with vertigo. Prospective studies with large patient cohorts are required to determine a definitive treatment protocol and prognostic factors in children with SSNHL.

Conflict of interest: The authors declare no conflict of interest.

There are no financial conflicts of interest to disclose.

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