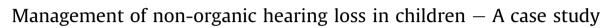
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ABSTRACT

A 10 year-old girl was admitted due to the claim of progressively developing hearing loss. The impedance audiometry showed no abnormalities but it was impossible to obtain reliable outcomes during pure tone audiometry assessment. The girl was additionally sent for speech audiometry, indicating a bilateral hearing loss and objective evaluations such as distortion product otoacoustic emissions and auditory brainstem responses, which results indicated a normal hearing. On the second day, repeated subjective audiometric tests showed also normal hearing, despite constantly reported hearing loss. After the psychological consultation and exclusion of neurologic pathology, the diagnosis of non-organic hearing loss was stated and the girl was scheduled for regular appointments with psychologist.

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1. Introduction

The extensive research on different types of ear-nose-throat (ENT) diseases contributed notably to our increased knowledge of characteristic outcomes found in different audiological entities and facilitated the diagnostic process. However, for many audiologists especially those starting their medical practice -a real problem arises when there is no consistency upon audiological evaluation. There are many possible reasons taken into consideration while obtaining conflicting results. One of the most complex issues is the hearing loss of non-organic origin. Its main audiological feature described in the literature is reporting a greater degree of hearing loss than found upon audiometric evaluation or situation when subjective hearing tests are indicative of much poorer findings than objective ones [1–6]. Many terms are used to describe this condition, such as "functional hearing loss", "hysterical hearing loss", "pseudohypacusis", "conversion hearing loss", "dissociative deafness", "simulated hearing loss", "malingering", "feigning", "psychogenic deafness" or "non-organic hearing loss (NOHL)" [7]. This diverse nomenclature reflects the variety of approaches aiming to explain the underlying cause of the disease, but at the same time it presents the lack of standardization in diagnostic process. As the

term "non-organic hearing loss" seems to be one of the most commonly used words and is considered by us also the most neutral in meaning, to provide a clarity we decided to utilize this term upon the whole article, after exclusion of any organic hearing loss origin.

Although there is a very limited literature concerning pediatric NOHL, most authors are in agreement that the prevalence of this condition can be underestimated, mainly due to the high misdiagnosis possibility [2,3,5]. Results from a study conducted by loannis at al. indicated that NOHL can be even the most common etiology of sudden hearing loss in children [6].

The accurate diagnosis of NOHL is particularly important. The underlying cause, if unrecognized, leads to needless prolongation of suffering and significant expenses such as unnecessary medical examinations or hearing aids fittings [1]. Psychological trauma, family and social conflicts are often found in children diagnosed with NOHL. For this reason, not only the participation of specialists such as audiologists, psychologists, psychiatrists, neurologists, but also involvement of family members is demanded in the most sufficient, interdisciplinary management strategy. The aim of our study was to present such an approach in a case study of 10-year old girl who came due to the hearing loss complaint.



Case Report



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2. Case report

A 10 year-old girl was admitted due to the claim of progressively developing hearing loss. The child's father reported that the deterioration was evident particularly in the last two months. The mother was absent during the visit because of health problems resulting in 6-week hospital stay. The girl started school education a year earlier than her peers and was attending fourth class of the mainstream primary school at the moment of otologic evaluation. Although lack of learning problems, the child was very ambitious and worried about her performance. Also, she was still mourning after the death of her grandmother, which happened six months earlier. The only health problem noted during the interview was skin allergy, but no medication was taken on the regular basis. Despite reported hearing loss, special attention of the audiologist aroused when the child responded to quiet commands without looking at the speaker's face and demonstrated no considerable communication problems.

The ENT examination revealed no pathological changes. The girl was sent for audiological evaluation, which included basic audiometric tests, i.e. tympanometry, acoustic reflex thresholds measurement and pure-tone audiometry (PTA). The impedance revealed no abnormalities. However, it was impossible to obtain reliable outcomes during PTA assessment. The experienced technician stated that there were great discrepancies among girl's responses (reaching 50 dB HL on average), even though she demonstrated complete understanding of conducted test's procedure. The decision about expanding the diagnostic protocol was taken and the girl was additionally sent for speech audiometry and distortion product otoacoustic emissions (DPOAE) examinations. In the study we used the Pruszewicz monosyllabic words speech audiometry. According to this standardized test used in our country, people with normal hearing obtain a full speech discrimination (100%) for about 50 dB SPL. Speech audiometry, if considered separately, revealed abnormal results indicating existence of significant hearing loss (Fig. 1). The girl was also scheduled for the auditory brainstem responses (ABR) evaluation. The ABR testing was conducted to exclude retrocochlear pathology. For this reason we decided to incorporate into diagnostic process 1000 Hz and click testing.

Normal results were obtained with DPOAE measurements. ABR were recorded using both tone (1 kHz) and click (2–4 kHz) and reached 20 dB nHL for both ears. The next day the physician explained to the patient and her father that the examination results were unclear and trying to perform PTA and speech audiometry once again was suggested. The girl was gently instructed that her cooperation is a basis of obtaining adequate results and she should respond as precisely and accurately as she could. She expressed the will to do so. The outcomes of repeated measures are shown in Fig. 2 (PTA) and Fig. 3 (speech audiometry).

On a basis of all the abovementioned examinations no organic cause of reported hearing loss was found. The girl was discharged in a good general state. She was commissioned for an appointment with a psychologist and simultaneously for neurologic examination. The neurologic examination was conducted to confirm our suspicion of the non-organic cause of the reported hearing loss and exclude the the nervous system pathology. During the next visit one week later, a psychologist stated that the emotional situation of the girl was unstable and she demanded regular psychological care at her residence place. The results of the neurologic examination from the reference specialists revealed no pathological findings favoring the suspicion of non-organic hearing loss origin. As the involvement of family members is a significant part of NOHL management, the child's father was informed that no organic cause of the reported HL was detected and the audiometric evaluations indicated a normal state of the auditory pathway. It was explained that the death of the grandmother and the separation from the mother could significantly influence the emotional state of the girl and result in the reported hearing loss problems. The father was also instructed that the problem should not be considered as a cheat attempt, but as a valuable communicate of a child needing help from the caregivers. Appointments with a psychologist were recommended to support the child and the family in dealing with this difficult event.

3. Discussion

Our short literature review on NOHL in children revealed that only 216 cases were documented in the literature since 2005 [1–6,8,9]. It is still not clear if this situation results from the lack of awareness among clinicians - leading to a high misdiagnosis percentage - or from the rarity of the complaint. Holenweg and Kompis reported that in a six-year period between 2003 and 2008, only 0,1% children out of 19,353 people admitted in general to their clinic were diagnosed with NOHL [1]. On the contrary, in two times longer observation time, children who matched the audiological NOHL criteria constituted 1,8% of the whole population undergoing ABR assessment in a study conducted by Schmidt et al. [3]. Drouillard et al. stated that in a shorted period of six months, even 6,2% of all evaluated children were diagnosed as suffering from NOHL. As we can see, data available nowadays is inconsistent and does not allow for making reliable assumptions concerning NOHL incidence.

What we can be more confident about is a pronounced preponderance of girls than boys diagnosed with NOHL, ranging from 61,5%-70% [1-4,6]. Interestingly, case reports found in the literature presented exclusively evaluation of boys with unilateral hearing loss [8,9]. To our best knowledge, it is the first article in the last ten years containing detailed NOHL assessment in a girl with deterioration in both ears. Regarding laterality of hearing loss, most authors are in agreement that bilateral cases are more common than unilateral, affecting 61% to even 94% of patients [1-3]. The only exception is the study of Ioannis et al., where unilateral NOHL was experienced by 65% of children [6].

Our patient was 10 year old at the time of audiological evaluation. Manifestation of NOHL in this period of time is compatible with findings from other studies, suggesting the peak incidence in children between 9 and 11 years of age.

Although children diagnosed with NOHL are not experiencing any serious physical health problems, relevant concomitant circumstances can be identified by the experienced audiologist during an interview and raise a special attention from the beginning of the diagnostic process. The most common are earlier experience with hearing loss (affecting the child or somebody from its environment) as well as school or home problems such as learning difficulties, physical aggression, abuse, death in the family, parent's separation or behavioral disorders [1,4]. At least three of the abovementioned situations were transparent in the case of the evaluated girl, i.e. death of the grandmother, longer separation with the mother and problems with adapting to demanding school conditions. No accompanying symptoms such as otalgia, tinnitus or vertigo, found in other hearing loss entities are reported by patients with NOHL [10] – such situation happened also in our patient's case as the girl reported exclusively progressive hearing deterioration.

One of the most significant principles in the field of audiometric testing is the cross-checking method introduced by Jerger & Hayes in 1976 [11,12]. This method emphasizes the importance of using appropriate, selected hearing tests and comparing their results in order to obtain the most accurate diagnosis. Such an approach is widely applied in the diagnosis of NOHL, where the need of

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