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REVIEW PAPER

Sudden sensorineural hearing loss in children – literature review

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ABSTRACT

Many cases of childhood sudden sensorineural hearing loss (SSNHL) remain debatable and cause both diagnostic and treatment problems. The aim of the study is to recall and collect the latest information about the problem of sudden hearing loss in children.

Sudden sensorineural hearing loss in children under 15 years old is rare and its pathophysiology remains unclear. Due to this fact, children's SSNHL's clinical characteristics, etiology, treatment outcomes and prognosis have been most likely deemed to be less relevant. There are many factors in children that are able to induce SSNHL, the main ones being viruses, autoimmune and genetic factors.

Examination includes tympanometry, audiometry, brainstem responses, as well as radiological testing and laboratory tests. The main treatment applied in this condition is corticosteroids. Prognosis is not promising because nearly half of affected children do not recover.

KEY WORDS:

D-dimer, children, urticaria, biomarkers, C-reactive protein.

INTRODUCTION

Sudden sensorineural hearing loss (SSNHL) is bilateral or unilateral sensorineural hearing loss with at least a 30dB decrease in threshold in three contiguous test frequencies occurring over 72 hours or less [1]. The age of SSNHL occurrence is mainly 25–60 years, with the range 46–49 years being the most common [2]. It has been reported that the incidence in adolescents and children is approximately 3.5% of the adult incidence. That shows that sudden hearing loss in children under 15 years old is rare and its pathophysiology remains unclear [3]. Due to this fact, children's SSNHL's clinical characteristics, etiology, treatment outcomes and prognosis have been deemed to be probably less relevant [4]. Categories of potentially identifiable factors have been distinguished, i.e. infectious (HSV, cytomegalovirus, etc.), autoimmune or systemic diseases (systemic lupus erythematosus – SLE, Wegener's granulomatosis, Cogan's syndrome, etc.), genetic, trauma, vascular diseases and tumors (acoustic neuroma, multiple myeloma, etc.) [1]. Currently, there are no specific tests or diagnostic criteria for children with SSNHL caused by viral infections, autoimmune diseases, systemic diseases, vascular diseases or metabolic diseases [4]. However, in the pediatric population, 20% with hearing loss have abnormalities on radiologic testing; however, it has not been determined whether computed tomography (CT) or magnetic resonance imaging (MRI) is a better modality for SSNHL [5]. Results of the research carried out with 87 children and 707 adults diagnosed with sudden deafness from September 2003 and August 2012 showed that degree of hearing loss, gender, side, and recovery rate were similar in children and adults, but the rate of complete recovery was higher in children (60.0% vs. 45.4%, $p < 0.05$) [6].

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MATERIAL AND METHODS

A PubMed search was conducted using the keywords "SSNHL" "Children," and "Deafness." A total of 72 papers were found, from which 23 were selected as sources for the following discussion. UpToDate was searched for keywords "sudden sensorineural hearing loss" and "hearing loss in children". In the selection of publications the size of the study group and the year of publication were important; also the number of citations of the article was significant.

EPIDEMIOLOGY

Sudden sensorineural hearing loss in children is not common, and approximately 6.6–10% of reported cases of sudden sensorineural hearing loss occur in children under 18 years of age [7]; the mean age of patients reporting is 11 years [2]. Results of the research carried out with 75 patients showed that SSNHL occurs as often on the right as on the left, with a tendency to right-sided localization (53.33%) and bilateral hearing loss is only 4–19.9% [7]. Both in boys and girls, the incidence was very similar.

Another study found that prevalence estimates of hearing loss in school-aged children (around 6 years of age) range from 10/1000 to 15/1000 for mild bilateral hearing loss and 30/1000 to 56/1000 for unilateral hearing loss [8]. The results of research carried out on 1218 children (students of 3rd, 6th and 9th grades) showed that 5.4% of the study sample had minimal sensorineural hearing loss (MSHL). The prevalence of all types of hearing impairment was 11.3% [9]. Another study carried out between 1988 and 1994 with 6166 children aged 6 to 19 years showed that 14.9% of them had had low-frequency or high-frequency hearing loss of at least the 16-dB hearing level, 7.1% had low-frequency hearing loss of at least the 16-dB hearing level, and 12.7% had high-frequency hearing loss of at least the 16-dB hearing level. Most of the hearing loss was unilateral and concerned the 16–25-dB hearing level. Of those with measured hearing loss, 10.8% were reported to have current hearing loss during the interview [10].

ETIOLOGY

The number of diseases where sudden idiopathic deafness may occur continues to increase along with the emerging etiological and environmental factors. Categories of potentially identifiable factors have been distinguished, i.e. infectious, autoimmune, genetic, traumatic and vascular.

The etiology in children is ambiguous; however, as the research shows, it is most often associated with an earlier viral infection; unlike adults with a dominant microcirculation defect, viral etiology accounts for 25% of SSNHL cases [5] and they are: cytomegalovirus (CMV), which is also the cause of SSNHL in 0.2–2.5% of liveborn newborns [11], Epstein-Barr virus (EBV), herpes simplex virus (HSV) type 1 and 2, mumps, rhinovirus, and SARS-CoV-2.

Autoimmune factors account for about 5% of SSNHL cases, one of which is autoimmune inner ear disease (AIED), which accounts for 4–30% of cases; bilateral and asymmetric incidence predominates, more often in girls with an average age of 8–9 years. The cause is not fully understood, but it is a good prognostic factor as it responds well to steroid and immunosuppressive therapy. 85.7% of patients responded to the treatment. A useful diagnostic marker is the anti-HSP-70 protein [12].

Less common autoimmune diseases include SLE, juvenile rheumatoid arthritis, juvenile rheumatoid arthritis as well as Cogan syndrome and Wegener's granulomatosis.

A genetic link was also demonstrated; 21.4% of patients reported the presence of SSNHL in a family where it was most frequent in siblings, 38.9% [13]. In a meta-analysis of 47 studies, 26 genes were selected from among 68 genes. Most of the studied genes are related to thrombosis, the inflammatory response, and oxidative stress or free-radical damage. The selection of such genes is based on the fact that the presence of thrombosis, oxidative stress, and inflammation plays an important pathogenic role in development of SSNHL [14]. However, the influence of genes, such as methylenetetrahydrofolate reductase, protein kinase C, complement factor H15, and lymphotoxin alpha, on SSNHL has already been reported [15].

Among the trauma and deformity factors correlated with SSNHL most frequently reported in the literature are enlarged vestibular aqueduct syndrome (4–8%), Mondini dysplasia (3%), absent/hypoplastic cochlear nerve (5%), acoustic trauma (4%), and fracture of temporal bone (2%) [16]; causes include lesions in the labyrinth, internal auditory canal, cerebellopontine angle, brainstem, and cerebral cortex.

DIAGNOSTICS

Examination of children suffering from sensorineural hearing loss (SHL) should include otomicroscopy and formal audiology, radiology and laboratory tests. Formal audiology helps to differentiate where the abnormalities are located [1]. In children, audiologic assessment that requires little cooperation is preferred. Tympanometry and specific forms of audiometry are performed (visual reinforcement audiometry, play audiometry). Brainstem responses are, however, the most important objective examination. They can differentiate between conductive, cochlear and neurologic hearing conditions. Otoacoustic emissions can help in examining cochlear function [17]. It is helpful to screen for widespread infections that are known to cause hearing disorders, as well as markers of autoimmune diseases. As many as 34.3% of children with SSNHL can have anti-CMV antibodies, 23.2% HSV and 20.2% rubella antibodies, but it was not determined

whether hearing loss is the delayed onset of a congenital disease or acquired infection and whether hearing loss is related to infection [18].

In adults MRI is a preferable radiology test. It should be conducted within 3 months of symptoms' onset [19]. In children it is still under debate whether MRI or CT is a better option. Computed tomography appears to be more helpful in imaging anatomical abnormalities. Magnetic resonance imaging is however very helpful in imaging neoplasms, such as cerebellopontine angle tumors. In imaging, 24.2% of adolescent patients present abnormal findings [18].

TREATMENT AND REHABILITATION

Sudden sensorineural hearing loss (SSHL) in children is relatively uncommon in comparison to that in adults. For this reason, successful treatment in this age group is still under debate. Some cases of children's SSHL appear to represent laboratory and imaging findings that can help to choose the most suited treatment [20]. The role of vaccines against *Haemophilus influenzae* type B (HiB) and *Streptococcus pneumoniae* in preventing SHL should be emphasized [1], as well as avoiding ototoxic drugs. Antiviral drugs should not be administered. Efficacy of applying them has not been proven; it is hard to measure their antibody titers. In cases of pediatric Idiopathic sudden sensorineural hearing loss (ISSHL), the main drugs to be applied are corticosteroids. Effectiveness of systemic and combined systemic and intratympanic injection treatment in children is similar [20], with the suggestion that combined therapy might have better results [21]. In adults most recent studies show that implementation of intratympanic corticoid and prolonged orally administered corticoid is the most effective treatment. Dexamethasone and prednisone are used [22]. Because of a lack of recommendations and data about treatment of pediatric ISSHL, treatment resembling that in adults should be applied [20]. There is a lack of evidence of effectiveness of other, nonsteroidal medication in children. Some drugs used in adults can be implemented. In adults hyperbaric oxygen therapy can have positive results. However, in adults it is used in cases resistant to steroids due to high cost [19]. Antiviral drugs, thrombolytics and pentoxifylline are not recommended [21, 23]. Good prognostic signs are unilateral hearing loss, tinnitus and ascending audiogram. Also age > 12 is considered as beneficial in children although in adult patients older age is less profitable. Poor prognostic signs are profound hearing loss and delay of treatment > 6 days. About 46.7% of children do not recover despite treatment [18].

DISCUSSION

Sudden sensorineural hearing loss is a serious problem and its treatment should be considered as a matter of emergency. Still little is known about its etiology, treatment and prognostic factors in children [7]. Sudden sensorineural hearing loss is a major diagnostic challenge in children who have not developed speech yet, because they are not able to convey symptoms, which can significantly delay the start of treatment. It is often the case when the child does not cooperate in the diagnostic process; it can be influenced by many factors, such as an inappropriate attitude of the doctor or negative experiences of the child and prejudices during previous visits to the doctor. The solution to these problems may be the education of parents in the field of observation and hearing tests in the period after the acquired pathogens, which are the main etiological factors. On the other hand, the education of doctors in dealing with uncooperative patients regardless of age, as well as communication, plays an increasingly important role in the accuracy of diagnosis and the shortest possible time interval for starting treatment.

The results of research carried out with 1218 children (students of the 3rd, 6th and 9th grades) showed that 5.4% of the study sample had MSHL [9]. Tarshish et al. reported the incidence of bilateral hearing loss in 45% of children who took part in the research (9/20) [24]. Na et al. reported that 8.1% cases of hearing loss in children were of the bilateral type [6]. In Yi's research 72 children (96%) had SSNHL in one ear, including 40 children (53.33%) in the right ear, 32 (42.67%) in the left ear, and three children (4.00%) in both ears [7]. Hence youth may have a problem with learning outcomes during SSNHL, because they cannot attend lessons and this reduces contact with peers; also longer hospital stays and prolonged treatment, often without significant improvement, can reduce quality of life.

CONCLUSIONS

Sudden sensorineural hearing loss is rare in children, with viral infections (CMV, EBV, HSV type 1 and 2, SARS-CoV-2, etc.) and autoimmune diseases being the main factors, while in adults microcirculation defect is the main cause. In addition to hearing examination, MRI should be conducted to exclude neoplasms. The most commonly used is steroid therapy. Due to the lack of literature, further research on the effectiveness and way of administration of the therapy is necessary. Considering that recommended therapy in adults is combined intratympanic and systemic steroid therapy, such treatment can be administered to children. The approach to every patient should be individual. Some patients do not respond to treatment; other treatment possibilities may be unavailable. Antiviral drugs, thrombolytics and pentoxifylline should not be administered because the efficacy of their application has not been proven. Hyperbaric oxygen therapy can be considered. We recommend further research on treatment options.

DISCLOSURE

The authors declare no conflicts of interest.

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