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Sudden sensorineural hearing loss

Nagły niedosłuch czuciowo-nerwowy

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Keywords

sudden sensorineural hearing loss, sudden deafness

Summary

Sudden sensorineural hearing loss (SSNHL), commonly known as sudden deafness, is an increase of bone and air conduction thresholds of greater than or equal to 30 decibeles affecting at least three consecutive frequencies within a 72-hour window. The loss of hearing is commonly accompanied by tinnitus and, rarely, vertigo. The vast majority of SSNHL is of unknown cause. Even 32 to 65% of cases of SSNHL may recover spontaneously. Treatment includes systemic and topical steroids and hyperbaric oxygen therapy (HBOT). Antivirals, thrombolytics, vasodilators, or vasoactive substances should not be routinely prescribed. Patients who have local neurologic findings require computed tomography scan of a head. A retrocochlear pathology should be excluded by obtaining magnetic resonance imaging or auditory brainstem response. The follow-up audiometric evaluation should be obtained at the conclusion of treatment and within 6 months of completion of treatment. The presentation of SSNHL in pediatric population is commonly delayed. Treatment options are similar to those of adults but the hearing recovery rate is higher.

Słowa kluczowe

nagły niedosłuch czuciowo-nerwowy, nagła głuchota

Streszczenie

Nagły niedosłuch czuciowo-nerwowy (SSNHL), powszechnie zwany nagłą głuchotą, definiuje się jako podwyższenie progu słyszenia krzywej kostnej i krzywej powietrznej o co najmniej 30 decybeli dla trzech kolejnych częstotliwości w przeciągu 72 godzin. Utrata słuchu jest często związana z pojawieniem się szumów usznych lub, rzadziej, zawrotów głowy o charakterze vertigo. Etiologia większości przypadków SSNHL jest nieznana. Nawet 32-65% pacjentów doświadcza spontanicznej poprawy. Leczenie obejmuje sterydoterapię (systemową i miejscową), a także terapię tlenem hiperbarycznym (HBOT). Leki antywirusowe, trombolityczne, wazodylatacyjne czy wazoaktywne nie powinny być przepisywane rutynowo. Jedynie w przypadku stwierdzenia ogniskowych zaburzeń neurologicznych powinna zostać wykonana tomografia komputerowa głowy. Poza tym należy zawsze wykluczyć patologię pozaślimakową na podstawie skanów rezonansu magnetycznego głowy czy słuchowych potencjałów wywołanych. Kontrolne badania audiometryczne powinny być wykonane tuż po i 6 miesięcy po zakończeniu leczenia. W populacji pediatrycznej postawienie rozpoznania SSNHL jest trudne, często opóźnione, ale rokowanie co do poprawy słuchu lepsze niż w populacji osób dorosłych.

Introduction

Sudden hearing loss (SHL) is defined as a rapid-onset subjective sensation of hearing impairment in one or both ears. The hearing loss may be either conductive, sensorineural or mixed. Sudden sensorineural hearing loss (SSNHL) affects in Poland 5 to 20 per 100 000 adults annually and is sometimes referred colloquially as "nerve hearing loss" (1, 2). The true incidence in pediatric population is unknown. It is estimated to occur in approximately 3-10% of all cases (3). The audiometric criterion for the entity is an increase of bone and air conduction thresholds of greater than or equal to 30 decibels affecting at least three consecutive frequencies within a 72-hour window. Patients may complain of ear fullness prior to the SSNHL onset. Tinnitus and vertigo may be co-occurring symptoms. Tinnitus, when persistent, may become a primary concern. Vertigo, on the other hand, is associated with poorer prognosis for hearing recovery in adults (1, 2). The vast majority of cases of SSNHL affects only one ear. Bilateral cases have been reported to occur in < 2% of adults and are associated with poorer prognosis (4-6).

Ethiology

SSNHL indicates the abnormal functioning of the cochlea, auditory nerve or even central nervous system. It may be caused by pathologies in cerebello-pontine angle (a.o. vestibular schwannoma), stroke, malignancy, noise, ototoxic medications, trauma. Nevertheless 90% of cases is considered to be idiopathic (2, 7-10). Causes of SSNHL in most of patients is never identified. Possible etiologies include infection, central nervous system pathology, inner ear pathology, autoimmune disease, vascular compromise. Viral infections include: herpes simplex virus, varicella zoster virus, human immunodeficiency virus. Bacterial pathogens which may influence development of SSNHL are: mycoplasma, Lyme disease, tuberculosis, syphilis (1, 2). An association of autoimmune disease with SSNHL has been noted by several studies (11-13). Cerebral microangiopathy co-occuring with diabetes, hypertension and hyperlipidemia in older patients could be associated with worse prognosis. Nevertheless, there is insufficient evidence that any routine laboratory test will impact the diagnosis, treatment or prognosis (2). Signs and symptoms which may suggest non-idiopathic SSNHL are presented in table 1.

Tab. 1. Signs and symptoms which may suggest non-idiopathic SSNHL

Sudden onset of bilateral hearing loss (Cogan's syndrome, viral infections, ototoxic medications, trauma, lead poisoning, genetic disorders, mitochondrial disorders, MELAS, sarcoidosis, hyperviscosity syndrome)
Antecedent fluctuating hearing loss on one or both sides (Meniere's disease, Cogan's syndrome, hyperviscosity syndrome)
Concurrent severe bilateral vestibular loss with oscillopsia
Gaze evoked or downbeat nystagmus
Concurrent eye pain, redness, lacrimation and photophobia
Focal neurologic symptoms or signs: headache, confusion, diplopia, dysarthria, focal weakness, focal numbness, ataxia, facial weakness (stroke, brainstem and auditory cortex infarct, anterior inferior cerebellar artery occlusion, severe atherosclerotic narrowing of the vertebrobasilar vessels, multiple sclerosis, meningitis, neoplasms)
Recent head trauma
Recent acoustic trauma
Recent barotrauma

Management and prognosis

Even 32 to 65% of cases of SSNHL in adults may recover spontaneously (2, 14, 15). Treatment include systemic and topical steroids and hyperbaric oxygen therapy (HBOT). Other options mentioned in the literature are as follows: antiviral agents, rheologic agents, diuretics, other medications, herbal treatments. The presence of fistula requires middle ear surgery. The alternative is observation alone (2).

Newest clinical practice guidelines for sudden hearing loss include 13 key action statements (KAS) introduced in 2019 by American Academy of Otolaryngology-Head and Neck Surgery Foundation (2):

1. Clinicians should distinguish sensorineural hearing loss from conductive hearing loss when the patients first presents with SHL.
2. Clinicians should assess patients with presumptive SSNHL through history and physical examination for bilateral SHL, recurrent episodes of SHL, and/or focal neurologic findings.
3. Clinicians should not order routine computed tomography (CT) of the head in the initial evaluation of patients with presumptive SSNHL.
4. In patient with SHL, clinicians should obtain, or refer to a clinician who can obtain, audiometry as soon as possible (within 14 days of symptom onset) to confirm diagnosis of SSNHL.
5. Clinicians should not obtain routine laboratory tests in patients with SSNHL.
6. Clinicians should evaluate patients with SSNHL for retrocochlear pathology by obtaining magnetic resonance imaging (MRI) or auditory brainstem response (ABR).
7. Clinicians should evaluate patients with SSNHL about the natural history of the condition, the benefits and risks of medical interventions, and the limitations of existing evidence regarding efficacy.

8. Clinicians may offer corticosteroids as initial therapy to patients with SSNHL within 2 weeks of symptom onset.
9a. Clinicians may offer, or refer to a clinician who can offer, hyperbaric oxygen therapy (HBOT) combined with steroids therapy within 2 week of onset of SSNHL.
9b. Clinicians may offer, or refer to a clinician who can offer, HBOT combined with steroid therapy as salvage within 1 month of onset of SSNHL.
10. Clinicians should offer, or refer to a clinician who can offer, intratympanic steroid therapy when patients have incomplete recovery from SSNHL 2 to 6 weeks after onset of symptoms.
11. Clinicians should not routinely prescribe antivirals, thrombolytics, vasodilators, or vasoactive substances to patients with SSNHL.
12. Clinicians should obtain follow-up audiometric evaluation for patient with SSNHL at the conclusion of treatment and within 6 months of completion of treatment.
13. Clinicians should counsel patients with SSNHL who have residual hearing loss and/or tinnitus about the possible benefits of audiological rehabilitation and other supportive measures.

Complete anamnesis and physical examination should distinguish SSNHL from conductive cases. The patient should answer the question about prior trauma, ear pain, ear canal instrumentation, ear drainage, fever, neurologic symptoms, ear fullness, tinnitus, vertigo. Physical examination includes otoscopy with inspection of ear canals and visualization of tympanic membrane. Certain pathologies (cerumen impaction, otitis media with effusion, acute otitis media, foreign bodies, external otitis, trauma, cholesteatoma etc.) can be excluded at this point. Weber and Rinne tests (tuning forks 256 or 512 Hz) are helpful in distinguishing types of hearing loss when otoscopy revealed no pathology. If the both tests are consistent with each other, the sensitivity may reach even 95% (16). Weber test could be unreliable in more than 20% of cases. When Weber alone lateralizes away from the worse hearing ear, the sensitivity may reach 99% (17). Those results should prompt the audiometric testing with appropriate masking, which should be performed within 14 days of symptoms onset (2).

Patients who may require a CT scan have focal neurologic findings, a history of trauma, chronic ear disease. CT should be considered in patients with known bone disease (a.o. Paget's disease, fibrous dysplasia, metastasis to temporal bone). It should not be routinely performed in patients with SHL in emergency departments. In fact, routine CT scan of the head should be considered unnecessarily harmful due to the radiation dose of 1-10 mSv (2).

MRI is the gold standard for excluding pathologies co-existing with SSNHL. While it is extremely sensitive and widely available, it is costly and risky in terms of gadolinium administration (immediate reactions to gadolinium, gadolinium-induced nephrogenic systemic fibrosis, deposition of gadolinium-based contrast agent in the brain) (18, 19). The overall intracranial incidental findings on brain MRI are common. The rate of pathogenic MRI abnormalities directly related to the SSNHL ranges from 4.4 to 13.75% (20-25). Those are direct causes of SSNHL (cerebello-pontine angle tumor, cochlear inflammation, multiple sclerosis) or findings that imply an underlying etiology (small vessels cerebral ischemia). Retrocochlear pathology should be excluded based on MRI scans, ABR, and in certain cases – temporal bone CT scans. Cerebellopontine angle tumors, represented in majority of cases by vestibular schwannoma, have a relatively high prevalence (2.7-10.2%) and be the first symptom in patients with SSNHL (9, 22, 26-28). Associated events of disease such as barotrauma or recent viral infection are present in one third of patients with vestibular schwannoma. Hearing recovery does not exclude the tumor (9, 22).

Nevertheless, there are situations in which MRI scans cannot be obtained (a.o. patients with peacemakers, severe claustrophobia, those, who do not wish to have MRI). It should be replaced by contrast-enhanced temporal bone CT scans or ABR. Normal ABR does not exclude retrocochlear pathology and may miss an average of 20% of intracranial vestibular schwannoma tumors (18, 29, 30). It is highly sensitive for tumors greater than 1cm in size and those in cerebellopontine angle. However, it may not be reliable, if hearing threshold exceeds 80 dB at 4000 Hz (31, 32).

The treatment includes immediate systemic (IV or oral) and/or intratympanic (IT) corticosteroids therapy. The treatment should be implemented within 14 days of SSNHL onset and the full dose should be continued for 7 to 14 days and then taper over the similar time period. Combination therapy (systemic + IT) may improve hearing outcome up to 13 dB (33). Contraindications for systemic corticosteroids therapy are certain pathologies such as: insulin-dependent or poorly controlled diabetes, labile hypertension, glaucoma, tuberculosis, peptic ulcer disease, prior psychiatric reactions to corticosteroids, pregnancy. This group should receive IT steroid injections. IT injections should be offered in case of incomplete recovery 2-6 weeks after initial systemic corticosteroid therapy (2).

HBOT exposes a patient to 100% oxygen at a pressure level 1.5-2.0 atmosphere absolute in a specially designed chamber. This facilitates delivery of oxygen to the cochlea, which is very sensitive to ischemia. HBOT can be offered by clinician as a primary therapy additional option within 14 days of initial therapy (2).

A salvage therapy including systemic or IT corticosteroid therapy and/or HBOT can be offered within one month of symptoms onset. Up to date none of the studies reported statistically significant benefit from antiviral, rheologic or vasoactive agents (2). Follow-up should be obtained for 6 months after completion of treatment.

SSNHL in pediatric population

The presentation of SSNHL in pediatric population is commonly delayed on average from 8.6 to even 122 days (3, 34). Wood et al. (3) conducted a meta-analysis study of management and treatment outcomes in patients under 19 years old. Based on 13 studies (605 patients, 670 ears) they found out that 24.2% of children had abnormal findings in CT or MRI scans of head. The most common serological finding was cytomegalovirus immunoglobulin (IgG or IgM) and constituted 34.3% of ears tested. Bilateral SSNHL occurred in 25.4% pediatric patients. Treatment includes systemic and intratympanic steroids and HBOT – same as in adult population. The partial/total improvement of hearing was associated with following factors: unilateral hearing loss, tinnitus, age > 12 years, ascending audiogram. On the other hand, decreased odds of improvement were associated with: profound and/or bilateral hearing loss, delay of treatment > 6 days. The steroid therapy (general or local) had no impact on increased odds of partial or complete improvement. The meta-analysis does not support vertigo as a negative prognostic factor. Partial improvement or complete resolution of treatment was reported for 53.3% ears. The authors divided SSNHL pediatric population into two groups: older children (> 6 years) with truly idiopathic etiology who may have similar prognosis to that of adults, and younger children with underlaying congenital disease (genetic, anatomic) or immunologic cause of hearing loss. The latter are less likely to experience recovery of hearing.

Conclusions

SSNHL is a relatively rare disease which requires an urgent treatment. The loss of hearing is commonly accompanied by tinnitus and, rarely, vertigo. The vast majority of SSNHL is of unknown cause. Treatment includes systemic and topical steroids and might be complemented by HBOT. Even 32 to 65% of cases of SSNHL may recover spontaneously. It is important to exclude retrocochlear pathology based on MRI scans, auditory brainstem response (ABR), and in certain cases – temporal bone CT scans. The follow-up audiometric evaluation should be obtained at the conclusion of treatment and within 6 months of completion of treatment. The diagnosis of SSNHL in children is often delayed. Treatment options are similar to those of adults but the hearing recovery rate is higher.

Conflict of interest
Konflikt interesów

None
Brak konfliktu interesów

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