Unilateral Sensorineural Hearing Loss (USNHL): Still a Challenge to Manage

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Abstract

Unilateral sensorineural hearing loss (USNHL) is a type of hearing impairment where there is normal hearing in one ear and impaired hearing in theother ear. Patients with unilateral hearing loss have difficulty inhearing conversation on their impaired side, localizing sound, understanding speech in the presence of background noise, interpersonal and social relations. Usually, it is felt that patients with severe-to-profound USNHL function normally. It is now well established, however, that USNHL is a handicap that can negatively impact QOL. Limited literature is available regarding its incidence, psychological impact and treatment. Hence, it is a challenge to manage a patient with USNHL.

Keywords: Sensorineural Hearing Loss; CROS; SNHL; Unilateral.

Introduction

Hearing impairment cannot be seen and hence its effects are not visible to others, so deaf suffers in silence. Unlike blindness deafness often provokes ridicules rather than sympathy [1]. Imagine pressing your ear with one hand and trying to hear with the other. In unilateral sensorineural hearing loss (USNHL), hearing loss is present only in one ear. The other ear will be having normal hearing capacity. It can be just a mild hearing loss, that does not affect daily functioning or it can be a very major loss with a severe or profound intensity, causing hearing handicap. When a patient can hear from only one ear, and there are limited possibilities to compensate for the handicap, e.g., changing listening position, group discussions and dynamic listening situations become difficult. Individuals with profound unilateral hearing loss are often perceived as socially awkward due to constant attempts to maximize hearing leading to socially unique body language and mannerisms [2]. Unilateral hearing loss is found to be occurring in both children and adults. People with unilateral hearing loss are found to have difficulty in distinguishing sound and speech from the background noise.

Incidence of USNHL

There are very few studies on incidence of USNHL.

Hearing loss is an extremely common disorder, with approximately 32 million Americans having some degree of impairment. Sensorineural hearing loss (SNHL), which accounts for 90 percent of the cases, is caused by damage to the cochlea or the vestibule-cochlear nerve. The vast majority of patients with SNHL have bilateral hearing loss. In the United States, approximately 60,000 new cases of USNHL occur annually (www. singlesideddeafness.com) and far more occur internationally [3].

In an Indian retrospective study done on audiometric data of 252 patients to know the demographic characteristics of patients suffering from hearing loss- 163(66.5%) had Sensorineural hearing loss (SNHL), out of which only 6 cases (3.7%) had unilateral SNHL[4].

Another Indian study on 820 patients diagnosed with pure sensorineural hearing loss (SNHL) on Pure Tone audiometry, 675(82.3%) had bilateral involvement and unilateral (USNHL) was in 145 (17.6%) patients. Amongst unilateral cases, right ear involvement was in 48.2% cases and left ear involvement was in 51.8%[5].

Causes of Unilateral Hearing Loss

Hearing loss can occur suddenly, wiping out hearing within 72 hours (Sudden Sensorineural Hearing Loss- SSNHL), and it can affect anyone of any age.Interestingly, most cases of sudden onset hearing loss are single-sided. Sensorineural unilateral hearing loss, affecting both the nerves and the inner ear, is irreversible.

It is difficult to find out the root cause for the occurrence of unilateral hearing loss in a particular case. But the general causes for the occurrence can be classified as detailed below.

- 1. Trauma to the ear or parts of the ear
- 2. Exposure of the ear to excessive and continuous noise
- 3. Genetic hearing loss
- 4. Infections and illnesses of various types

Numerous disease processes can lead to severeto-profound USNHL. These include Congenital, sudden SNHL; idiopathic SNHL; neoplasms; vestibular schwannoma (acoustic neuroma); demyelinating pathologies such as multiple sclerosis, vertebrobasilar arterial occlusion (stroke), acoustic trauma, head injury, perilymphatic fistula, ototoxic drugs, labyrinthitis, Meniere's disease; and autoimmune disease (Cogan disease, Wegener's granulomatosis, lupus, Takayasu arteritis, systemic sclerosis, and other rheumatological disorders)[3].

What Happens in USNHL

Our physiology is designed for bilateral hearing, located on either side of the head. This design, with a space between the two ears, has several evolutionary benefits. The handicap experienced by adults with bilateral sensorineural hearing loss is well known, but the consequences of unilateral sensorineural hearing loss (USNHL) is often underestimated based on the assumption that a person with normal hearing in the contralateral ear is not likely to face a major handicap.

First, there is the notion of space which gives us the advantage of spatial hearing. When we hear a sound, the nerves of the ear closer to the sound gets stimulated just a little earlier than the ear on the other side. Just a micro-second of difference, but it helps the ear to determine which side the sound is coming from. This advantage gets even more pronounced with localization of hearing which helps us discriminate between sounds coming from a distance and sounds close by. The stereo sound effect, letting us hear sounds coming from 360 degrees, gives that rich full sound we are used to.Binaural hearing is vital for sound localization, speech discrimination in a background of noise, ability to identify common sounds and ease of listening. When these sensitive functioning get thrown off-radar, normal life can get quite confusing. For example, the person may not be able to understand normal speech, the direction of a sound, or from how far the sound is coming. This can cause accidents or hamper movement in regular life, while crossing roads, at home or in the workplace. With speech and background noise presented at the same level, persons with unilateral deafness were found to hear only about 30-35% of the conversation [6].

Usually, it is felt that patients with severe-toprofound USNHL function normally. It is now well established, however, that USNHL is a handicap that can negatively impact QOL [3]. A study of USNHL among Indian patients showed mild to moderate psychosocial handicap usingHearing Handicap Inventory for Adults (HHIA), in about a third of patients and no handicap in a little over half the patients. Sound localization and speech in noise are significantly affected in these patients. However, despite this, most patients do not choose to go in for any kind of rehabilitative device and present to the physician to be reassured that there is nothing sinister about their problem [7].

USNHL is Known to Cause: [8]

- Irritability
- Body language and mannerisms which appear socially awkward or unusual
- Frequent headaches, stress
- Social isolation
- Chronic interpersonal communication difficulties due to inability of brain to isolate or beam form sounds and voices of other individuals
- Appearance of anxiousness even in low noise situations

- Jumpiness
- Trouble figuring out where sounds are coming from.
- Trouble paying attention to what people are saying: "evasive" behaviour.
- Misdiagnoses as ADHD
- Seeming lack of awareness of other people's personal space and moods since brain is hyper-focused on deciphering auditory information in lieu of non-verbal social cues.
- Lack of sound depth: any background noise (in the room, in the car) is flat and wrongly interpreted by the brain. The effect is similar to what happens when trying to hear someone speaking in a noisy crowd on a mono TV. The effect is also similar to talking on the phone to someone who is in a noisy environment
- Inability to filter out background noise or selectively listen to only the important portion of the noise in the environment.
- For sensorineural hearing loss, the lack of input coming from the damaged sensory apparatus can cause "ghost beeps" or ringing/tinnitus as the brain attempts to interpret the now missing

Types of CROS Hearing Aids- [9], [10]

sensory data. The frequency and the volume of the noise can increase according to one's physical condition (stress, fatigue, etc.). This can aggravate social problems and increase the difficulty of speech comprehension.

 Talking loudly or "broadcasting": the affected person cannot perceive the volume of his or her voice relative to other people in the same room or close company, resulting in being characterized by others (who may be located beyond normal auditory range) as domineering or boorish.

Treatment

Learning of the central nervous system by "plasticity" or biological maturation over time does not improve the performance of monaural listening [8]. In case where surgical remedy has been ruled out by an otolaryngologist, then the following types of amplification of sound signals is the probable method of treatment.

Contralateral Routing of Signals (CROS) hearing aids Hearing aids that take sound from the ear with poorer hearing and transmit to the ear with better hearing.

Conventional CROS/ Bi CROS	comprises a microphone placed near the impaired ear and an amplifier (hearing aid) near the normal ear. The two units are connected either by a wire behind the neck or by wireless transmission. The aid appears as two behind-the-ear hearing aids and are sometimes incorporated into eyeglasses.
	<u>CROS</u> -is for a user who has relatively normal hearing in the good side and has hearing that can't be aided on the bad side. The receiving BTE device on the bad side transmits the sound to a device on the good side. The user hears the amplified sound from the bad side in their good ear.
	<u>BICROS</u> - is for a user with little or no hearing on one side and with some hearing loss in their better ear. It works just like the CROS implemenation, except that the device on the good side is actually a fully capable hearing aid for hearing sounds from the good side that is also capable of receiving the sound transmitted from the CROS aid on the other side.
CIC transcranial CROS	comprises a bone conduction hearing aid completely in the ear canal (CIC). A high-power conventional air conduction hearing aid fits deeply into the patient's deaf ear. Vibration of the bony walls of the ear canal and middle ear stimulates the normal ear by means of bone conduction through the skull.
BAHA transcranial CROS	Bone Anchored Hearing Aid (BAHA): a surgically implanted abutment transmits sound from the deaf ear by direct bone conduction and stimulates the cochlea of the normal hearing ear.
Sound Bite Intraoral bone conduction	which uses bone conduction via the teeth. One component resembles a conventional behind- the-ear hearing aid that wirelessly connects to a second component worn in the mouth that resembles a conventional dental appliance.[8]
Cochlear implant has also been rec	commended in cases of USNHL (severe- profound) with intractable tinnitus. [11]

There have been very few studies comparing CROS systems. One study of the BAHA system showed a benefit depending on the patient's transcranial attenuation [11]. Another study showed that sound localisation was not improved, but the effect of the head shadow was reduced [12].

Conclusion

Limited work has been published on incidence and causes of Unilateral SNHL. The functional and psychological impact of USNHL is underestimated. Limited options are available to help and treat such patients. The usage adaptability and cost constraints of CROS hearing aid is a challenge to manage patients of USNHL. Further studies and research is required in the field of USNHL to know the incidence, functional and psychological impact and treatment options.

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