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Mini Review

Current Perspectives in Management of Unilateral Sensorineural Hearing Loss and Single Sided Deafness

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ABSTRACT

Unilateral Sensorineural Hearing Loss (USNHL) or even Single Sided Deafness (SSD) were mistakenly believed in the past that they could not induce a notable negative effect on the average individual adult. Respectively, a child with USNHL could eventually develop typically and adequately with no particular challenges. Today, it is well established that both children and adults with USNHL and SSD experience difficulties locating sound sources than their normal peers attributable to the concomitant deprivation of data utilized for localization; interaural time differences along with interaural intensity differences, especially for high frequency sounds. Moreover, USNHL and SSD patients suffer from the absence of the binaural benefits that permit people with bilateral Normal Hearing (NH) to perform relatively well in challenging listening environments. These benefits encompass binaural summation that causes improved speech perception, and binaural release from masking that facilitates word recognition in noise. Rising treatment strategies, involving various type of amplification, Assistive Listening Devices (ALSs) and Cochlear Implantation, can greatly widen our overall approach regarding USNHL and / or SSD. Nevertheless, most recent evidence points out that both prompt and adequate intervention is crucial to promote optimal outcomes.

Keywords: *Single sided deafness; Unilateral hearing loss.*

INTRODUCTION

Unilateral Sensorineural Hearing Loss (USNHL) is defined as the particular (sensorineural) type of Hearing Loss (HL) that affects only one ear (with audiometric thresholds >15db regarding children and >20db regarding adults). On the other hand, Single Sided Deafness (SSD) refers to the condition where one ear has normal pure tone sensitivity whereas the lateral one has nonfunctional hearing.[1]

The conventional approach in the past regarding USNHL was that it could not induce a notable negative effect on the average individual adult and that a child with USNHL could eventually develop typically and adequately, with no particular challenges.[1] However, there has been strong evidence since 1980s indicating that young patients suffering from USNHL do exhibit developmental, academic, social, and behavioral deficits, far more often in comparison to their own peers.[1]

Nowadays, it is well established that although such a unilaterally impaired child seems to function quite efficiently in everyday classroom situations, there's a great amount of evidence that actually suggests quite the opposite. Leaving a permanent HL untreated can become a persistent long-term problem therefore strong effort should be made to compensate for it, at least in the classroom environment. Among children, USNHL definitely needs to be treated as a top priority.[2] Similarly, among adults, parallel rules apply with the distinctiveness that each adult's own lifestyle and situational needs should also need to be taken into consideration.[1,2]

That being the case, it's the health professional's responsibility to interact with the family, with the adult or the child who is the patient, and with the available support services, whether it's the school system itself or otherwise, in order to provide them with the optimal

available intervention options.

IMPACT OF USNHL AND SSD ON CHILDREN AND ADULTS

Both children and adults with USNHL and SSD experience bigger trouble locating sound sources than their normal peers attributable to the concomitant deprivation of data utilized for localization; interaural time differences that specify data for low frequency sounds, and interaural intensity differences respectively for high frequency sounds.[1,3] It is worth mentioning the fact that both age groups are cognizant of the impediment they experience regarding sound localization.[1]

Moreover, USNHL and SSD patients suffer from the absence of the binaural benefits that permit people with bilateral Normal Hearing (NH) to perform relatively well in challenging listening environments (either when background noise is present or when there is intense reverberation). These benefits encompass binaural summation that causes improved speech perception, and binaural release from masking that facilitates word recognition in noise.[1] Furthermore, the head shadow effect can additionally impart to not perceiving high frequency consonants, that are crucial for speech perception, particularly when the noise is pointed towards the 'good' ear and the signal of interest towards the ear with the hearing deficit.[1,3] In order to perform equally well, patients with USNHL or SSD need a a more clear Signal to Noise Ratio (SNR) of more than 2 dB even in the more favorable listening condition.[1]

In terms of academic performance, educational and pedagogical accomplishments of children with USNHL or SSD have been analyzed in multiple ways encompassing grade holding, requirement of special services, and teacher own reports.[1] As evidence suggests, 25%-50% of children with USNHL and SSD undergo academic rigorousness involving grade retention and the necessity of special education services due to difficulties experienced inside the school environment.[1] Moreover, although there was a strong belief in the past that these patients were not supposed to present any speech and language developmental draw backs, or difficulties in language competence, (due to the auditory input to the normal side), this is not the case. Recent evidence suggests that up to 40% of children with USNHL and SSD are in danger for worsen speech and language abilities than their peers, with symmetrical hearing competence, although they may catch up later over time.[1]

As far as social and behavioral development is concerned, current research suggests that up to one third of children with USNHL or SSD exhibit difficulties in the social and behavioral field, and that emotional consequences to the encountered challenges, derived of the unilateral hearing deficit, continue well into adulthood.[1,3] With regard to cognitive development, findings have been variable and inadequate so far, although there are some indications that children with USNHL or SSD may have lower performance on verbal and functioning intelligence quotient measures in comparison to their normal peers.[1] More recent studies consistently show older students with asymmetrical hearing loss to perform considerably worse than their peers in both mathematics and language skills.[2]

Generally, current evidence suggests that children with USNHL or SSD are a dissimilar group of patients and the effect of hearing impairment is versatile and often unpredictable. However, a major part of these children does tend to exhibit difficulties in multiple areas, in comparison to NH children.[1] Interestingly enough, the degree of the difficulties can be further differentiated due to factors unrelated to the deficit itself, such as family's own socio-economic potential and mother's education level.[1,4]

TREATMENT BASICS

A plethora of management choices exist, especially for patients with USNHL / SSD. They incorporate among others classroom modifications (when indicated), amplification, Contralateral Routing of Signal hearing aids (CROS), Bone-Anchored Hearing Aids (BAHA), Cochlear Implants (CI) and Assistive Listening Devices (ALSs).[1] It should be mentioned that various interventions may be suitable for each individual.[1] Any intervention protocol must be tailor made taking into consideration not only the patient's own particularities but also his / her family's own resources, needs, and expectations.

Initial and long-term counseling regarding the possible course and the consequences of the unilateral hearing impairment effects and the crucial significance of early management, is vital to ensure that patients and their families make informed choices. Targeted counseling can emphasize in the necessity to safeguard the NH ear from loud noise along with routine supervision for potential middle ear infections.[1] Such conditions may significantly impact the individual with unilateral sensorineural hearing deficit who strongly relies on his / her NH ear.

CONVENTIONAL HEARING AID

Evidence indicates that a conventional Hearing Aid (HA), in cases of USNHL, can help patients achieve better listening sensation along with improved performance in terms of academic and social conditions. Moreover, utilizing the HA unilaterally, aided patients with USNHL to experience greater comfort regarding listening in quiet and noise even though there were no subjectively detectable alterations in speech recognition to support this.[1] These findings were further supported by parents', teachers', and children's own reports mentioning meaningful HA gains at home, inside classroom, plus in overall quality of life.[1,5]

Toddlers with mild to severe USNHL have also exhibited much better performance in terms of localization, besides the fact that when they were fitted later they did experience bilateral interference.[1] Surprisingly enough, only one quarter of children with USNHL use their HA all day long; compared to three quarters of children with a moderate HL affecting both sides.[1] This reduced HA utilization may be due to the typically poorer compliance generally observed in children fitted with HA later in their lives.[1,6] Further evidence necessitates with respect to the potentially best pediatric prescriptive fitting method for USNHL as it demands active patient engagement and therefore is far from adequately optimized for fitting infants and uncooperative toddlers.[1]

Adults with USNHL also tend to obtain positive outcomes with conventional HA usage. The primary positive predictors for HA utilization in adults were societal and job demands - activities along with digital signal processing availability. This finding suggests that although individual requirements on communication may affect type and functions of HA choice, high technology features may best serve grown-ups with USNHL.[1]

The bottom line is that any sufferer from asymmetric hearing loss should be considered as a candidate for a HA fit. Each patient

should at least go through a HA trial to experience its potential benefits in an adequate time frame.[7]

CONTRALATERAL ROUTING OF SIGNALS (CROS) HEARING AID

A CROS HA functions by placing a microphone on the affected ear that consequently transmits sound through amplifier and receiver to the contralateral NH ear. The receiver is usually coupled to an earmold for proper retention.[8] A CROS HA is mainly indicated in cases of profound USNHL or SSD. The desired benefit derives from the fact that signals delivered towards the impaired ear can be successfully detected by the normal one.[2] More specifically, the main advantage of transmitting sound to the NH ear is the enhanced perception of high frequency signals from the damaged, contralateral side.[8] This relies on the fact that frequencies ≈ 2 KHz and above, coming towards the side of the auditory deficit, cannot be perceived by the healthy side due to the head shadow effect.[3]

Optimal results can be obtained when the patient is placed in a noisy background where noise reaching the normal ear is not aggravated.[1] That being said, CROS HAs are not suitable in the first place for young children who can't monitor or control their (noisy) environment in order to place themselves in an acoustically advantageous position on their own. In quiet situations, CROS HAs seem to be able to meliorate speech perception and localization, especially regarding adults.[1,2]

BONE-ANCHORED HEARING AID (BAHA) SYSTEM

A BAHA system is used typically in cases of conductive or mixed HL, but can be also appropriate for severe to profound USNHL or SSD. In older children and adolescents with USNHL or SSD, a bone anchored implant transmits sound from the opposite ear to the normal cochlea through bone conduction.[8] It can be surgically implanted in young patients aged 5 years and more, or alternatively placed with the help of a soft headband in younger ones.[1]

Evidence regarding BAHA systems in children is relatively recent although most of the children implanted between the ages of 6-16 years reported subjective improvement in various listening conditions and overall better quality of life.[6] Patient satisfaction was even better in cases with children suffering from conductive hearing losses and in cases of congenital unilateral atresia.[1,6]

Similar literature data also apply to adults, who have experienced improved auditory performance in difficult listening environments, as well as better quality of life in general.[1] Nevertheless it seems that BAHA provide better results in conductive rather than sensorineural type of impairments and that is why only about one third of patients with profound USNHL or SSD obtain long term advantages from a BAHA utilization.[1]

FREQUENCY MODULATED (FM) SYSTEM

FM systems wirelessly forward the sound of interest to either the hearing-impaired ear (when applicable) or the NH ear, in order to enhance Signal to Noise Ratio (SNR). They decrease by this way the negative effects of reverberation, potential distance from the sound source itself and noise.[1,7] Treatment of unilateral HL (especially in cases of children) with FM systems may raise pitfalls related to a vast range of potential options and growing evidence on targeted outcomes.[8] These systems may be utilized in various ways such as being coupled to the NH ear in cases of SSD or profound USNHL,[9] listener's own HA, at ear level in an open fitting, with headphones or ear buds, with a desktop speaker, or with loudspeakers all around the classroom.[1] The proper type of FM system implemented should be settled upon the specific child's needs, classrooms' specifications - infrastructures, and listening environment particularities.[1]

Recent evidence nevertheless suggests that FM systems enhance speech perception in children with USNHL both in quiet and noise. Surprisingly enough, improvement in speech perception is present even when the noise targets the impaired side.[1,7] As the magnitude of USNHL grows up, so does the FM benefit regarding speech recognition. This applies especially in cases of system with a non-occluding fitting, whose benefit is even superordinate, compared to that of the conventional or CROS HA fit.[1]

COCHLEAR IMPLANTS (CI)

CI provide immediate electrical stimulation to the auditory nerve, thus bypassing the cochlea, should this be the site of lesion in cases with profound USNHL or SSD.[7] With regards to adults' studies, CIs seem to improve speech recognition in quiet and noisy environments. Furthermore, better speech perception is documented under dichotic compared to monotic listening situations, implying successful brain plasticity regarding binaural integration of both acoustically and electrically transmitted sounds.[1] Surprisingly enough, the benefits of better speech recognition in noise have been found to continuously improve over time up to three years after the initial CI implantation.

Besides that, benefits of cochlear implantation in adults with USNHL or SSD additionally include enhanced sound localization, reduction of distress of severe tinnitus, subjective improvement of the comfort of listening and the overall quality of life. In addition, CIs have also been found to provide adult patients with profound USNHL or SSD with superior listening experience in comparison to either a BAHA system or a CROS HA.[1]

In cases of children, there is poor evidence supporting CI implantation in patients with profound USNHL or SSD concluding that evidence based criteria need to be further developed.[1] Preliminary results based on a small number of school aged participants with USNHL, do point out binaural hearing advantages for speech perception in noise, steadier localization of sounds and subjectively perceived listening comfort.[7,10] A review of the current knowledge on the potential advantages of a CI implantation in similar cases determined that young patients with acquired USNHL are most prone to obtain similar benefits from a CI as adults.[1,10] Nevertheless, there are multiple concerns regarding the final judgment to operate on young children with congenital or prelingual USNHL or SSD as the impact of the unilateral deficit may not become evident soon enough, before the young patient reaches school age.[1] Yet, outcomes from a CI may be not be as beneficial at later ages because of the auditory deprivation that can affect brain's own plasticity mechanisms.[1,10]

CONCLUSION

Besides the fact that sufferers from USNHL or SSD share the common symptom of hearing impairment in one of their ears (with auditory function within normal limits in the lateral side), the magnitude of the deficit in their affected side may play a crucial role in how the condition

is managed by a hearing health care provider.[2] A key reason for health care professionals being quick to properly intervene and begin managing the symptoms of USNHL and SSD has to do with the condition's effect on the patient's communicative, social, and academic development.[1,2]

Especially when we deal with congenital USNHL (where the loss is not detected a priori) HA fitting is far more demanding.[11] In such a case, potentially exposing the impaired ear to imprudent sound intensities leads to unwillingness or even denial to HA use. Since neural plasticity takes time to allow adequate binaural integration of auditory inputs, a longer period of time is mandatory to achieve the necessary fine tuning required to adjust and accept the HA fitting.[11]

In addition to early identification of the unilateral deficit, it is also crucial for health care clinicians to consider all available management and intervention options in order to substantially improve the development of patients with USNHL / SSD and help them reach their maximum potential.[1] The bottom line is that binaural input and adequate auditory stimulation should be considered as top priority to encourage development whenever residual hearing allows.[8] In cases of non-functional hearing, CROS can be considered as a viable way to offer audibility of sounds from the impaired side when cochlear implantation is not applicable.[8] Rerouting the signal from the debilitated side to the normal one decreases the negative effects of the acoustic head-shadow and enhances listening in noise when the signal of interest is located at the impaired side.[12] Nevertheless, it cannot restore access to binaural cues. CI on the other hand, may provide some better access to these cues, but by no means can result in a full restoration of binaural hearing competency.[12]

Future longitudinal studies will help determine the most effective options for treating USNHL and SSD in children and adults as the majority of patients desire some sort of intervention rather than no management at all.[13] Rising treatment strategies for profound USNHL or SSD, involving cochlear implantation, can greatly widen our overall approach in general, regarding these cases. Nevertheless, most recent intervention strategies point out that although young children with congenital USNHL do benefit from late amplification fitting in a one-to-one communication basis, they fail to do so in more demanding listening environments.[14]

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