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Single-Sided Deafness, Tinnitus and Cochlear Implants

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Abstract

Background: Single-sided deafness is a situation of profound hearing loss on one side, with normal hearing contralateral or near them (better than 40 dB HL). The only way to rehabilitate the side with single-sided deafness would be through the cochlear implant.

Objective: The aim of this study is to describe the outcomes of a cochlear implant in a case of single-sided deafness with tinnitus and perform a short literature review of this topic. Methods: Retrospective study of medical records of a patient who underwent surgery treatment for single-sided deafness and tinnitus with cochlear implant.

Conclusion: Cochlear implantation was an effective alternative for a patient with single- sided deafness, resulting in improvements in speech recognition and the tinnitus.

Keywords: Single-sided deafness; Cochlear implants; Tinnitus; Profound hearing loss.

Introduction

Unilateral hearing loss is defined as hearing loss on one side, ranging from moderate to profound, sensorineural, conductive or mixed sensorineural and conductive [1,2]. Single-sided deafness (SSD) is a situation of profound hearing loss on one side, with normal contralateral or proximal hearing (better than 40 dB HL) [1]. The SSD may be congenital, may have acquired factors associated, be progressive and may even occur suddenly [1,3].

The main etiologies related to SSD are ototoxicity, Ménière's disease, trauma, inner ear infection, otosclerosis, schawnnoma, circulatory/metabolic diseases, and genetic and other factors [3].

The real incidence of SSD is no clear in the literature, but for asymmetrical hearing loss are described rates ranging to 1 to 3% in children population and for unilateral hearing loss in newborns the rate are 0.5 at 1000 newborns, and increases with aging. In adults is described that there are 200 new cases per million people in world every year, and of those patients rate ranges from 50.00-60.000 in the United States and 9.000 in the United Kingdom [4-9].

Treatment currently consists of two forms of auditory rehabilitation: adaptable external device (CROS system) or anchored to the bone (BAHA system – Bone-Anchored Hearing Aid) [10]. Both treatments don't actually rehabilitate the deaf ear, but attenuate and mask the unilateral hearing loss using the patient's well side [10-13].

The only way to rehabilitate the side with SSD is with a cochlear implant (CI), which provides electrical stimulation of the ipsilateral cochlea and the auditory nerve. Several authors cite the IC as an excellent treatment option for SSD [14-19].

The aim of this study is to describe the outcomes of cochlear implant in a case of single-sided deafness with tinnitus and perform a literature review of this topic.

Materials and Methods

Retrospective study of medical records of a patient who underwent surgery for SSD and tinnitus with cochlear implant.

Audiometric Testing

Audiologic tests included impedance, tonal and vocal audiometry.

Speech perception tests: In the preoperative evaluation, the patient underwent a test of speech perception. This was based on a list of words and phrases, developed and adapted to Portuguese by Bevilacqua et al [22]. The patient underwent the tests with hearing aids in a quiet and peaceful place.

Postoperatively, the patient repeated the speech perception tests one year after CI activation. The tests were performed using the cochlear implant (CI). The same audiologist performed all pre-and postoperative tests.

The THI questionnaire (Tinnitus Handicap Inventory) was applied to evaluate tinnitus in the patient. This questionnaire contains 25 questions and is characterized by being numbered. The questions evaluate the emotional, functional and catastrophic aspects of tinnitus.

Ethics

The institutional review board approved this study and the subject gave written informed consent.

Case Report

Male patient, 32 years-old, reported tinnitus in the left ear that began for 2 years earlier. The tinnitus started after surgery for stapedectomy in the left ear, two years before. After this episode, the patient developed a concomitant tinnitus and deafness on the same side.

The patient had a history of surgery for conductive hearing loss, with air-bone gap in the left ear of approximately about 25 dB, with an abnormal tympanometry and good speech discrimination on the left side. He had never had abnormal otoscopy or other otological complaints.

The patient reported that tinnitus was a constant hum and whistle that worsened in the presence of noise. He also complained of poor speech recognition in noisy environments.

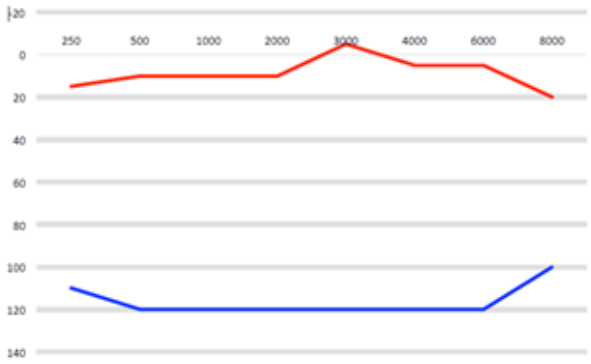

Figure 1: Tonal thresholds in dB NA in a pre-operative test. Blue= Left Ear; Red=Right Ear.

Speech perception – open set	Pre Op	Post Op (CI)
Recognition of sentences - S / N +10	50%	100%
Monosyllable recognition - S / N + 10	70%	100%
Recognition of sentences - S / N 0	70%	90%
Monosyllable recognition - S / N 0	80%	90%
Recognition of sentences - S / N - 10	50%	70%
Monosyllable recognition - S / N - 10	56%	70%
S=Sign/Sound, N= Noise; CI= Cochlear Implant on.		

Table 1: Evaluation of speech perception before and after cochlear implantation, with both ears.

The audiometric evaluation demonstrated cofosis in the left ear with absence of auditory brainstem response (ABR). On the contralateral side, there was no alteration (Figure 1). The assessment from imaging studies (CT and MRI) did not reveal any abnormalities with normal anatomy. The evaluation of his speech perception in the left ear is described in Table 1. The speech tests were normal in the right ear.

Regarding the tinnitus, the THI questionnaire (Tinnitus Handicap Inventory) was applied, and resulted in a score of 59. This result can be understood, since "Tinnitus promotes a functional interference and a difficulty to listen, creates confusion and worsens under stress" and it is classified as severe (grade 4 on a scale ranging from 1 to 5, where 5 is the worst).

The hearing aid test was not successful since there was no hearing improvement. The patient was evaluated using the Bone Anchored Hearing Aids (BAHA) Devices system, also for test. After an external test, the patient reported an improvement in sound perception and discrimination of speech. However, regarding his perception of tinnitus, there was no improvement.

After a short period, the factor that most bothered him was the tinnitus. Therefore, an evaluation process for a cochlear implant was initiated. His greatest expectations after the CI were improvements in the tinnitus and his speech recognition.

On CI activation, the patient's normal ear was occluded with a molded mass before beginning the device programming process. When the cochlear implant was first connected, the patient reported hearing "beeps" with no constant speech characteristics. After 5 minutes of use, the patient started hearing speech sounds.

After two months of CI use, the patient returned for follow-up and mappings. He arrived reporting that he already understood some words and his normal ear always helped him to understand when the implanted ear did not. He reported a significant improvement in tinnitus, saying that there were days when it was hardly noticeable.

The THI questionnaire (Tinnitus Handicap Inventory) had a great improvement, wit a 10 value obtained (Grade 1, that means "Slight: Only heard in quiet environment, very easily masked. No interference with sleep or daily activities"). It must emphasize that this index was 59 (grade 4 - severe) before CI surgery.

Free-field audiometry (position 45° with acoustic buffer on the normal side) was conducted two months after cochlear implantation, with almost normal pure tone thresholds in all frequencies achieved after CI surgery. (Figure 2)

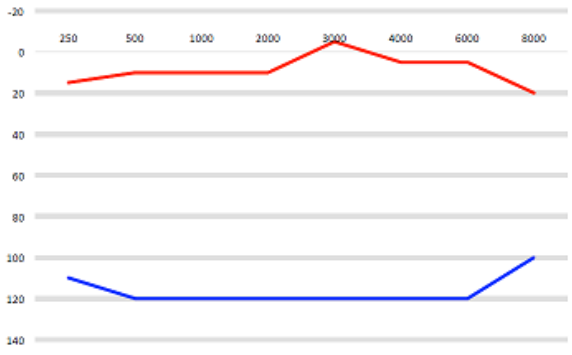

Figure 2: Free-field audiometry after cochlear implantation (2 months), in left ear (CI on).

The evaluation of the patient's speech perception in a postoperative test is shown in Table 1 (both ears) and 2 (left ear). It can be noted a huge improvement in speech outcomes, with both ears and also when comparing the left ear.

The patient was followed-up again 15 months after CI activation and there was no complaint of tinnitus at that time.

Speech perception – open set	Pre Op	Post Op (CI)
Recognition of nonsense syllables	0%	52.63%
Monosyllable recognition	0%	48%
Recognition of sentences	0%	74%
CI= Cochlear Implant on.		

Table 2: Evaluation of speech perception before and after cochlear implantation, with left ear only.

Discussion

SSD affects an individual's life in subtle ways due to adaptation to different listening situations using a unilateral hearing ear. Several specific mannerisms, such as where to sit, the position of the head, use of visual aids like lip reading and other similar habits, are common in such individuals. However, complaints and limitations are more evident in noisy environments, which decrease the intelligibility of speech, affect sound source localization, binaural squelch and head shadow effect, which can all impact the quality of life [10,11,14,20].

In the pediatric population, during the learning phase, the SSD can result in low educational attainment, poor language performance and speech deficits and introduce social difficulties such as low self-esteem and insecurity [11-13,21].

Research on tinnitus suppression by intracochlear electrical stimulation has gained interest over the past few decades and since the introduction of CI, it has become easier to apply [22].

Although individual studies on SSD and CI generally involve few patients and are low profile, but are shown that those subjects can achieve good improvements in sound localization and speech understanding in both quiet and noisy environments. In some cases, the outcome with a cochlear implant appears to be superior to those achieved with other devices, including the contralateral routing of sound devices and osseointegrated implants [23].

The cochlear implant is considered a safe procedure with a low rate of significant complications. A study on 403 patients who underwent cochlear implantation revealed a complication rate of only 19.9%, with the majority of complications being resolved by clinical management [24].

It is hypothesized that when the balance between excitatory and inhibitory action is broken within sensory systems, sensory circuits become plastic with adaptation to environmental inputs at synaptic levels. Tinnitus is an aberrant auditory sensation for all clinical types [25].

Clinically, it is considered as a reflection of failure in neuroplasticity, neuromodulations and neuroprotection in maintaining normal auditory functions at synaptic levels in the sensory cortex, which are projected to lower levels of the central auditory system in the brain and to sensorineural elements of the ear. Thus, cochlear implants are used to provide information to the affected side and decrease or eliminate the imbalance between the two ears in relation to stimuli reception [25].

A meta-analysis of 9 studies involving 36 patients with unilateral deafness who underwent CI, concluded that a CI for unilateral sudden hearing loss in patients with with a normal-functioning contralateral ear proved to be an effective therapy. Tinnitus is reduced, as is the signal-to-noise ratio, which allows 50% speech discrimination. All patients felt that they better localized sound, and most reported improvements in understanding speech [26].

A systematic review of 17 studies on 108 SSD patients undergoing cochlear implant concluded that they had improved speech perception, sound localization and tinnitus [27].

Several recent studies also observed improvements in speech understanding, sound localization and tinnitus. Because of the low number of cases, it is difficult to compare conclusively the outcomes achieved by cochlear implants with those achieved by other devices. However, on the basis of encouraging early results and its unique ability to restore binaural sound processing, a growing number of centers offer cochlear implant as a treatment for SSD [23,28].

The authors believe that CI should be implemented as early as possible in a case of sudden single sided deafness, and that CI is an excellent therapeutic option for patients with single-sided deafness and tinnitus, especially when the tinnitus appears after the deafness. However, we agree that the duration of deafness in patients with single-sided deafness is a controversial aspect for selecting CI as treatment.

Conclusion

We believe that CI is an effective option for a patient with SSD, with improvements in speech recognition and tinnitus, as reported in this patient's case.

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