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CASE REPORT

Cochlear Implant for Tinnitus and Hyperacusis: A Case Report

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Abstract: Tinnitus and hyperacusis are new challenged emergent indications of cochlear implantation. Some reports have demonstrated a suppression of tinnitus as a side effect after implantation. We describe the case of a 57-year-old man suffering from severe tinnitus and hyperacusis. Several years after onset of symptoms cochlear implantation was performed, which resulted in a progressive decrease of tinnitus and hyperacusis.

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Introduction

Indications of cochlear implantation are wide spreading through the field of auditory disorders and not only are recommended in severe to profound hearing loss. Some of these emergent indications of cochlear implantation are tinnitus and hyperacusis. Tinnitus consists of perception of sounds in the absence of an acoustic stimulus and it must be distinguished from the somato-sounds, which are sounds that are originated near the cochlea in vascular and musculoskeletal structures. Tinnitus is a prevalent symptom that affects from 15% to 30% of adult population [1].

In many occasions tinnitus is associated to hyperacusis which has been defined as consistently exaggerated or inappropriate responses to sounds that are neither threatening nor uncomfortably loud to typical person [2]. Hyperacusis may be distinguished from loudness recruitment that describes an experience commonly associated to cochlear hearing loss which consists of a perception of loudness level increases faster than normal with a rising sound level [3]. The prevalence of hyperacusis in patients with tinnitus as the primary complaint is about 40% [4] and the prevalence of tinnitus in the patients with a primary complaint of hyperacusis has been reported as 86% [5].

Tinnitus and hyperacusis could be very incapacitating in certain people. It is estimated that one to five per cent of the people who suffer from tinnitus are severely affected. Tinnitus may affect mental functioning and cause psychological distress, including anxiety and depression. There is great evidence that the annoyance of the tinnitus is not the perception of the tinnitus itself but the way in which patients experience the perception [6]. The same seems to be true in the case of hyperacusis. That is the reason why it is necessary to assess the grade of annoyance of both symptoms not only by an audiological assessment but by general and specific test to determine the impact in the quality of life too.

In the Ear Nose and Throat Department, Complejo Hospitalario Universitario Insular Materno Infantil de Gran Canaria, the people with severe tinnitus or hyperacusis are evaluated following an audiological protocol (including clinical interview, otoscopic examination, tympanometry and pure tone audiometry. As well, the patient undergoes a free field warble tone audiometry and disyllabic open words test [7] with and without hearing aids if patients used them.

As tinnitus is usually a subjective phenomenon, it is difficult to measure it using objective tests. Because of that we use tests such as comparison with noise of

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known frequency and intensity, as in an audiometric test. The objective is to assess the pitch, the loudness, the minimum masking level and the residual inhibition of the tinnitus.

Patients also have to complete the Tinnitus Handicap Inventory (THI) --Spanish version-- as well as the Sound Intolerance Questionnaire (SIQ) --Spanish version-- both of them validated by Herráiz et al. [8] [9].

The evaluation of tinnitus and hyperacusis is completed using two subjective scales, one for each symptom. A Subjective Discomfort Scale (SDS) is passed to the patients to assess the degree of discomfort caused by the tinnitus. Patients have to choose one of five possible situations: 1) If I do not pay attention I am not aware of tinnitus (without annoyance); 2) Tinnitus causes to me no problem all day, I notice it mainly if I sleep for a while (bearable); 3) It creates me problems in certain moments through the day (half nuisance); 4) It creates me problems all the day (continuing discomfort) and 5) It creates me problems all the day, tinnitus prevents me from leading a normal life (unbearable). In order to assess the annoyance caused by the hyperacusis, a Subjective Scale for Hyperacusis (SSH) is passed; the patients have to indicate the activities of normal life, which are affected by the intolerance to environmental sounds caused by the hyperacusis. There are ten activities in the list: 1) concerts; 2) restaurants; 3) cinema; 4) shopping; 5) social life; 6) go to church; 7) working; 8) driving; 9) sporting events and 10) take care of child. The higher the score in this test is, the greater the intolerance to sound, and so the degree of hyperacusis is higher.

Also, the severity of the annoyance caused by the tinnitus and the hyperacusis is measured by a Visual Analogue Scale (VAS) --in which 0 represents no discomfort and 10 the maximum discomfort--. Imaging tests and vestibular test could be done if the clinician considered it indicated.

Here we present the case of a patient with bilateral disabling tinnitus and hyperacusis who was treated successfully with cochlear implantation.

Case Presentation

A 57-year-old Asian man who has a Menière's disease of long evolution presented in our centre with complaints of severe disabling tinnitus. Pure tone audiometry testing revealed profound hearing loss in the right ear and severe hearing impairment on the contra-lateral side (pure tone average of 500, 1000 and 2000 Hz: 89 dBHL in the right ear and 70 dBHL in the left ear) (Figure 1).

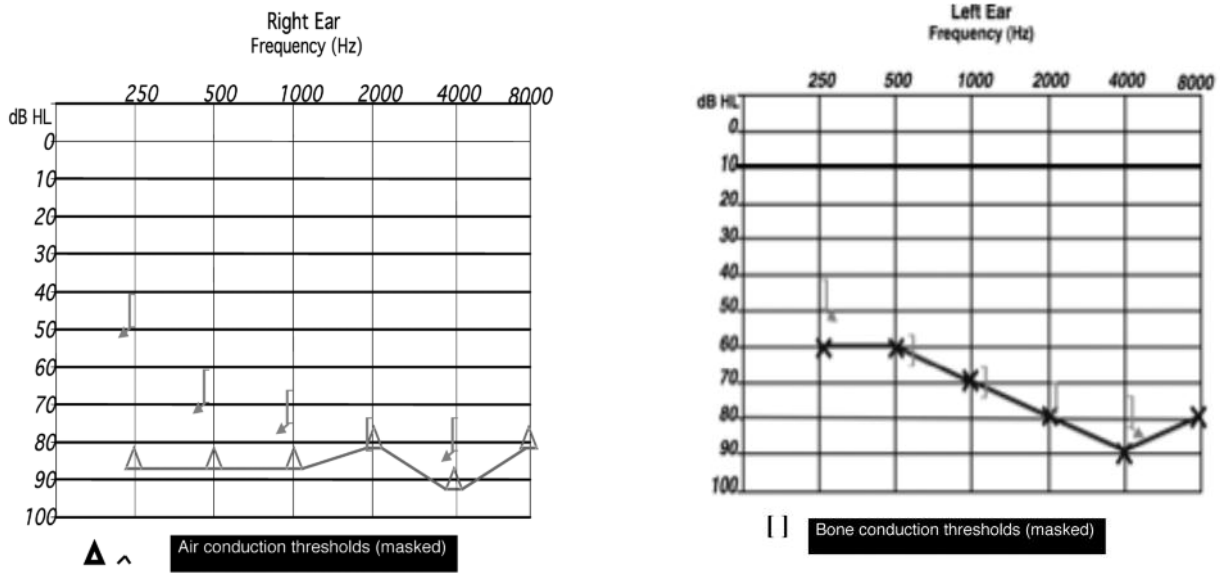

Figure 1. Hearing thresholds in April 2008.

The patient had a permanent tinnitus in the right ear, which had an intensity of 5 dBHL above the hearing threshold and a pitch near to a narrow band tone centralized at 250 Hz. In the left ear the patient had a fluctuant tinnitus, which was described to be as the sound of an engine. In the VAS he described his tinnitus annoyance as 4. In the SDS for tinnitus he marked a 3 (medium discomfort). The patient used a hearing aid since 2002 with a very poor result. In free-field condition tested with new adapted hearing aid, there was no speech perception in Korean language.

In the assessment of the hyperacusis the patient showed a great intolerance to any type of sound. He used to cover his right ear so any sound could annoy him. In the questionnaire of the hyperacusis disturbance he showed the maximum complaint.

In the caloric reflex test the patient showed a unilateral weakness of the 44% in the left ear and a directional preponderance of 46% in the right beating. The otoscopic examination was normal. The impedance test showed the stapes reflex at 110 dB SPL in three of the four frequencies tested (500, 1000 and 2000 Hz -- not at 4.000 Hz--) in the left ear and did not appear in the right ear. The preoperative Dissyllabic Open Word test, the THI and the SIQ could not be done due to the fact that the patient did not understand Spanish. The Magnetic Resonance Imaging and the Computed Tomography did not show any lesion in auditory pathways or in the inner ear.

The patient had been treated with Enoxaparin, potassium and sulpiride without any positive result, continued with vertigo attacks (more than two per month) and had tinnitus and hyperacusis all the time, so a labyrinthectomy followed by the location of an Advanced Bionics HiRes 90K® CI in the right ear was performed in May 2008. The surgical technique used was the transmastoid total labyrinthectomy with removing all of the vestibular neuroepithelium. No complication was observed. The function of the electrode array in the cochlea was evaluated by using intra-operative electrophysiological testing (Neural Response ImagingTM). There were no post-operative complications. One-month post implantation the patient underwent the first programming sessions (activation of the CI) in the context of a standard fitting procedure. During it, the audiologist adjusted program parameters (sound processing parameter values) performing a particular program or map.

The patient was followed for a period of time of 18 month after the implantation. He experimented an excellent evolution in his auditory perception, as he has an open set sentences test result in free field in Korean language of 85% using CD recordings at 65 dBHL without visual support and without repetitions, and he is learning Spanish with a good performance, and his tinnitus and hyperacusis decreased not only in the right side but also in the left side too. Figure 2 shows the excellent result obtained by the patient with the cochlear implant in free field conditions.

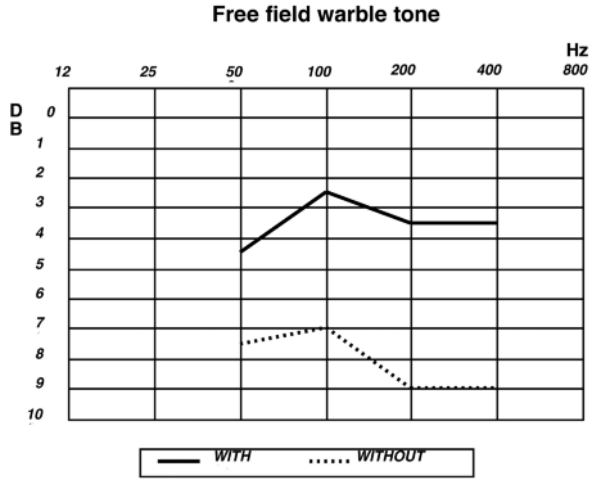

Figure 2. Free field warble tone audiogram (test conducted in attenuated room, with two loudspeakers located one meter away and 45º from the subject).

Figure 3 shows that the annoyance caused by the tinnitus (measured by the VAS and the SDS) decreased as well as the time of perception of the tinnitus. Eighteen months post implantation the patient perception of tinnitus decrease to only the 10% of the time and its severity had decreased so much that he referred its annoyance to be 1 in the VAS and 1 in the SDS.

The same happened to the hyperacusis. Figure 4 shows the decrease of the disturbance caused by the hyperacusis --measured by the VAS and the SSH--. Eighteen months post implantation the patient showed very low complaints of this hyperacusis (0 in the SSH – activities affected by the sound intolerance – and 1 in the VAS).

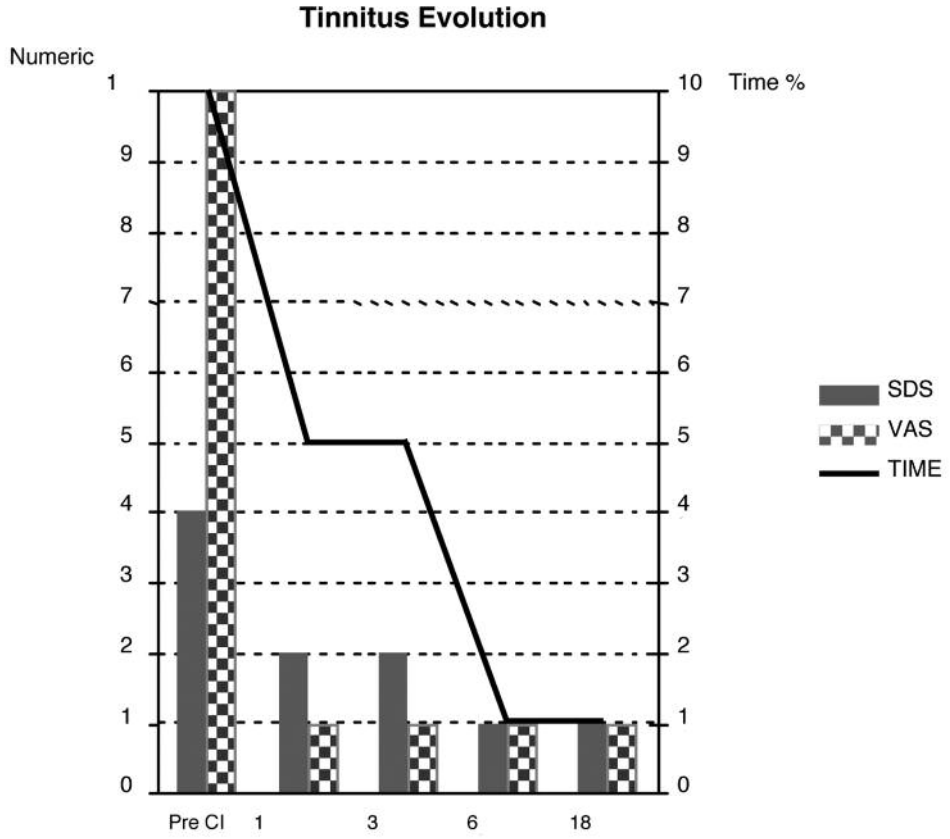

Figure 3. Evolution of tinnitus (18 months after implantation) SDS (Subjective Discomfort Scale); VAS (Visual Analogue Scale)

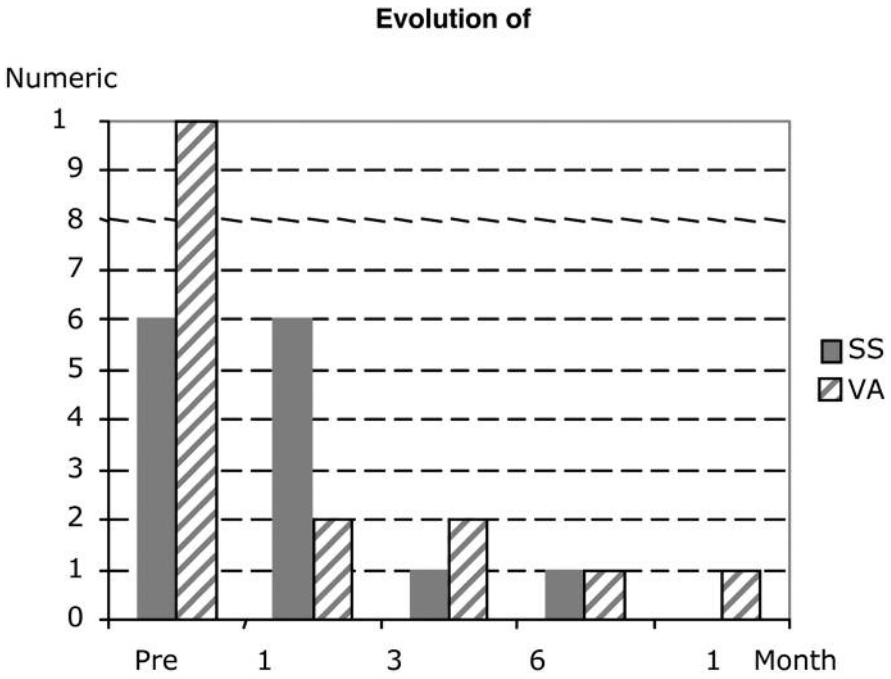

Figure 4. Evolution of hyperacusis (18 month post implantation). SSH (Subjective Scale for Hyperacusis); VAS (Visual Analogue Scale)

Regarding to the vertigo the surgery provided a good control of vertigo (Class A), the patient show improvement in imbalance and functional disability.

Discussion

Many treatments have been tested to reduce tinnitus and hyperacusis, however most of them are supported by a low evidence level or have suggested a limited benefit. One of the emergent treatments of these symptoms is the electrical stimulation of the cochlea by a cochlear implant. It is well known that the reduction of tinnitus after the implantation has been reported in several cases, even in the contra-lateral ear [10] [11] [12]. By means, pre-implanted tinnitus is reduced or suppressed in the cochlear implanted ear in 60 to 90% of the cases and in the contra-lateral ear in a similar rate. Post-implanted tinnitus or the worsening of pre-implanted tinnitus has been reported in less than 10% of cases.

Lusting et al. [13] evaluated the outcomes of cochlear implantation in 9 patients with long-term Menière's disease. They found a clear benefit in hearing, so they proposed the cochlear implant to treat the patients with Menière's disease who have developed a bilateral severe-to-profound sensorineural hearing loss. The same happened to our patient who showed a significant improvement in hearing. Almost all patients of Lusting's study had controlled vertigo symptoms. The most likely explanation of this fact is that most patients had arrived to the "end stage" of the disease progress in which vertigo attacks disappear and cochlear symptoms progress. However, this only happens in progressive forms of Menière's disease, which are estimated to be almost a quarter of all cases. Five of the nine patients of Lusting's study had undergone surgical procedures to control vestibular symptoms of Menière's disease. As our patient, one of them had a labyrinthectomy in the implanted ear, but it had been performed previously of the surgery of the cochlear implant. The transmastoid labyrinthectomy is the "gold standard" surgical technique to control vestibular symptoms in an ear with severe-to-profound hearing loss in the context of a Menière's disease. The subject who had undergone the labyrinthectomy previously to the cochlear implantation in Lusting's study did not perform as well as the other patients of that study in the audiometric test and continued having vertigo attacks during the first months after the surgery. However, after the third month after the surgery the patient's disabling vertigo had been controlled and he performed better in all audiometric tests.

The improvement of the tinnitus in our patient was better than the one the patient of the cited study had experimented because he had not vertigo attacks since the implantation. Thus, the labyrinthectomy has been successful to control vestibular symptoms and the cochlear implant has improved patient's hearing. Since Chen et al. [14] suggested that cochlear implantation could be done in a laryrinthectomized ear, there are many studies that confirm that cochlear implantation improves the sound awareness, speech recognition and communication in a previously labyrinthectomized ear. Thus, as Lusting et al. [13] suggest, cochlear implantation is a mechanism that will allow clinicians to be more aggressive in controlling disabling vertigo with interventions such as labyrinthectomy or intratimpanic gentamicine, which can worsen the hearing.

The hearing rehabilitation of the patients has been very complicated as well as the diagnostic because the patient did not speak Spanish before the surgery. The patient is currently following the programme of rehabilitation after cochlear implantation so he is learning Spanish as he improves his auditory and communications skills.

There are two basic theories that support the peripheral origin of tinnitus [15]: essentially it has been assumed that there is a chronic depolarization of the afferent auditory pathway by a mismatch between the functioning of the inner and outer hair cells or alterations in the neurotransmitter glutamate at peripheral level. Furthermore, in Menière's disease it is supposed that alterations in the homeostasis and pressure of the endolymph cause the chronic depolarization of the afferent auditory fibres.

However, neurectomy or labyrinthectomy do not cause the remission of the tinnitus in Menière's disease in all cases [16]. Therefore, it has been postulated that although the genesis of tinnitus may occur in the cochlea, the mechanisms that lead to the chronicity of tinnitus take place at the central nervous system.

Even in repose, the cochlea shows electrical activity spontaneously. Tyler et al. [17] propose that certain changes in the spontaneous activity of the cochlea are

the cause of the genesis of tinnitus. Therefore, the suppression of tinnitus may be possible simulating the baseline activity of the cochlea, and it could be done without perceiving the stimulation as Rubinstein et al. [18] proved by applying a high-rate pulsatile electrical stimulation to the cochlea in 11 subjects who suffer from disabling tinnitus (8 subjects reported the suppression of tinnitus and even 5 of them without perceiving the stimulation).

In most individuals, it is possible that the tinnitus originated at peripheral level is no longer perceived due to the adaptation to chronic stimulation. However, to Hallam et al. [19], there are some people who are unable to adapt to the tinnitus and, in these people, the "phantom auditory perception" persists indefinitely. Indeed, it is possible that the loss of tonic random afferent input may result in a loss of inhibition within brainstem auditory structures. One of these structures is the auditory efferent system. Therefore, it is possible to reduce or suppress hyperacusis by using a cochlear implant.

Moreover, other parts of the central nervous system are involved in tinnitus and hyperacusis generation and maintenance. It has been proposed that a maladaptive cortical reorganization after differentiation of the auditory cortex occurs is most individual after the onset of tinnitus [16]. Thus, the reorganization of the right auditory association cortex induced by the cochlear implant could reduce or suppress tinnitus even in bilateral cases as it has happened in our case. Our patient as well as others has reported a reduction or suppression of tinnitus even when the cochlear implant is switched off. The most reasonable explanation to that fact is that the changes induced by cochlear implant in auditory cortex and the residual inhibition, which consists of the reduction of tinnitus after a period of masking.

Cochlear implant a possible solution for patients with profound hearing loss and tinnitus. Further studies are needed to establish the indication of cochlear implantation in patients with tinnitus.

Conclusion

Tinnitus and hyperacusis are new challenged emergent indications of cochlear implantation. A patient with long-term Menière's disease with bilateral severe-to-profound hearing loss, frequent vertigo attacks, bilateral tinnitus and hyperacusis has been treated successfully in our Department by a cochlear implant. However, further studies are needed to establish the indication of cochlear implantation in patients with tinnitus and hyperacusis.

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