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Original Contribution

Round and oval window reinforcement for the treatment of hyperacusis

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ABSTRACT

Purpose: To present the outcomes of two patients (three ears) with hyperacusis treated with round and oval window reinforcement.

Materials and methods: Transcanal placement of temporalis fascia on the round window membrane and stapes footplate was performed. Loudness discomfort level testing was performed. Results of pre and post-operative hyperacusis questionnaires and audiometric testing were reviewed.

Results: Two patients (three ears) underwent surgery. Results from the hyperacusis questionnaire improved by 21 and 13 points, respectively. Except for a mild loss in the high frequencies, no change in hearing was noted post-operatively. Both patients reported no negative effects from surgery, marked improvement in ability to tolerate noise, and would recommend the procedure to others. There were no complications.

Conclusions: Round and oval window reinforcement is a minimally invasive option for treating hyperacusis when usual medical therapies fail. Further studies are needed to evaluate the effectiveness of the procedure in reducing noise intolerance.

1. Introduction

Hyperacusis, defined as noise intolerance to ordinary sounds, is a challenging problem encountered by otolaryngologists. Patients often present with emotional (anxiety, stress, depression), social (isolation, limitation in activities), and physical (pain, discomfort) symptoms. Individuals with hyperacusis suffer from a reduced quality of life, due to anxiety and noise-related avoidance in daily activities [1]. Although individuals with hyperacusis report varying degrees of intolerance to sound, the primary complaint is a physical and/or psychological reaction to sound. The physical properties of certain sounds elicit negative reactions in patients with hyperacusis which do not evoke an adverse reaction in an average listener [2]. The reported prevalence of hyperacusis ranges from 5.9% to 17.2%, depending on the definition of hyperacusis used and whether individuals with hearing loss are excluded in various studies [3,4].

The mechanism of hyperacusis is not completely understood, but has been related to acoustic overexposure resulting in increased gain within the central auditory pathways [5,6]. Contributing factors include a history of head trauma or acoustic trauma, hearing loss, and aging [7,8]. Hyperacusis is often accompanied by tinnitus and vice versa [9,10]. While tinnitus may arise from failure of the brain to adapt to deprived peripheral input, hyperacusis is thought to be related to an "over-adaptive" increase in response gain, as a result of afferent neuronal degeneration of auditory fibers [8].

Current commonly used treatment options for hyperacusis include avoidance of provocative stimuli, cognitive behavioral

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therapy, tinnitus retraining therapy, hearing amplification devices, and gradual sound exposure using wideband noise generators, with varied rates of success [2,11]. Although no surgical technique exists specifically for the treatment of hyperacusis, surgical intervention has been found to improve hyperacusis in patients with superior semicircular canal dehiscence (SSCD). Silverstein et al. found round window reinforcement to be an effective and minimally invasive surgical option for reducing the symptoms of SSCD [12,13]. Nikkar-Esfahani et al. noted an improvement in noise tolerance in patients with SSCD chiefly complaining of hyperacusis who underwent complete surgical occlusion of the round window [14]. Complete resolution of hyperacusis has also been reported in a case of unilateral posterior and superior canal dehiscence treated with transmastoid plugging of both defects [15].

The success of round window reinforcement in improving hyperacusis in patients with SSCD led the senior author to realize the potential benefits of performing a similar procedure in patients with a chief complaint of hyperacusis without evidence of SSCD. This paper reports the outcomes of two patients (three ears) with no evidence of SSCD, who underwent transcanal round and oval window reinforcement for the treatment of hyperacusis.

HYPERACUSIS QUESTIONNAIRE

*Please read each item below and rate it based on the Four-Point Scale.	0 NEVER Correct	1 SOMETIMES Correct	2 OFTEN Correct	3 ALWAYS Correct
1. Sounds that didn't disturb me earlier frighten me now.	☐	☐	☐	☐
2. I worry that I will never succeed in getting used to loud/uncomfortable sounds.	☐	☐	☐	☐
3. I cannot listen for a long time when I am surrounded by loud/uncomfortable sounds.	☐	☐	☐	☐
4. Because of my hypersensitivity to sound, there is tension between my partner and/or my family and myself.	☐	☐	☐	☐
5. I have to avoid certain sounds.	☐	☐	☐	☐
6. I am very scared of noise.	☐	☐	☐	☐
7. I think the hypersensitivity to sound has ruined my life.	☐	☐	☐	☐
8. When surrounded by a lot of sounds, I don't understand anything.	☐	☐	☐	☐
9. Other people distance themselves from me because I can't stand loud/uncomfortable sounds.	☐	☐	☐	☐
10. I am annoyed by sounds that are too loud/uncomfortable for me.	☐	☐	☐	☐
11. Loud/uncomfortable sounds cause physical pain in my ears.	☐	☐	☐	☐
12. I believe I won't be able to cope in everyday life if hypersensitivity to sound continues to be this bad.	☐	☐	☐	☐
13. I immediately withdraw when there are loud/uncomfortable sounds.	☐	☐	☐	☐
14. I am afraid that loud/uncomfortable sounds damage my hearing.	☐	☐	☐	☐
15. Since becoming hypersensitive to sound, I no longer enjoy music.	☐	☐	☐	☐

Date ________ Last Name ____________________ First Name ____________________

Fig. 1 – Validated hyperacusis questionnaire.

2. Methods

2.1. *Loudness discomfort level testing*

Loudness discomfort level (LDL) testing was performed after establishing pure tone thresholds at 250, 500, 1000, 2000, 3000, 4000, and 8000 Hz. Sound stimuli was presented starting at 60 dB HL and increased in increments of 5 dB HL. As the tone approaches the uncomfortable loud level, the step size is decreased in order to determine the LDL with a 1 dB resolution. This process is performed twice at each frequency, and the average of the two LDLs is recorded. The normal reference level for the LDL is traditionally accepted to be at 100 dB HL, although normal hearing individuals have been found to have LDLs between 86 and 98 dB HL [16].

The patients completed a validated hyperacusis questionnaire to rate the severity of their pre-operative and post-operative symptoms (Fig. 1) [17].

2.2. *Surgical technique*

Under general anesthesia, transcanal round window niche and oval window reinforcement was performed using a traditional transcanal tympanomeatal flap approach. If needed, the bony posterior canal was enlarged using a high-speed drill or curette to allow visualization of the ossicular chain, round window niche, chorda tympani, horizontal facial nerve, and hypotympanum. The bony lip overlying the round window niche was also removed with a one millimeter diamond drill for further exposure of the round window membrane. Temporalis fascia was obtained through a 2 cm incision above the auricle and small pieces were gently placed against the round window membrane and the stapes footplate (Figs. 2 and 3), and held in place with gelatin foam (Gelfoam). A 3 mm biopsy punch can be used to cut the fascia for the round window niche, and a 2 mm biopsy punch can be used to cut the fascia for placing over the stapes footplate. Following reinforcement, the tympanomeatal flap was repositioned and the external auditory canal was packed for one week with polyester packing strips and a small sponge.

2.3. *Patient 1*

A 64-year old male presented with a several-week history of positional vertigo consistent with benign paroxysmal positional vertigo. He was successfully treated with an Epley maneuver. On further questioning, he also reported an 18-year history of noise

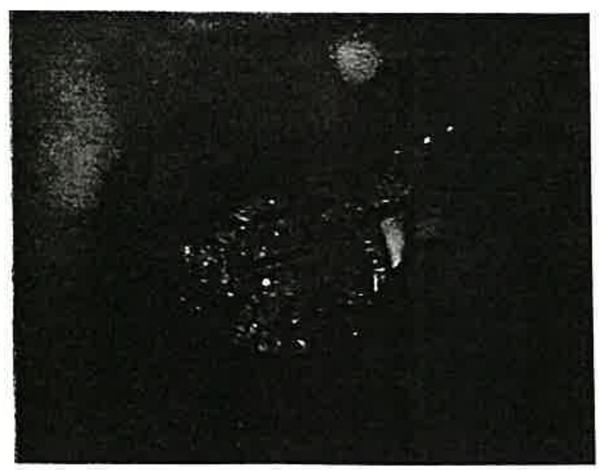

Fig. 2 – Left ear, transcanal approach. Temporalis fascia is covering the round window niche (small arrow) and the stapes footplate (big arrow). Incudostapedial joint (IS).

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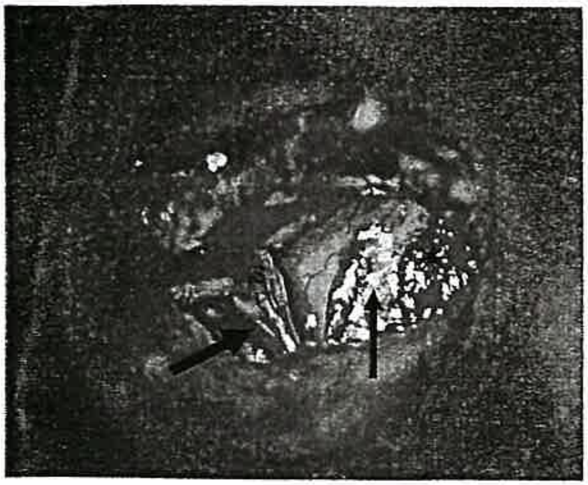

Fig. 3 – Right ear, transcanal approach. Temporalis fascia is covering the round window niche (small arrow) and the stapes footplate (big arrow). Gelfoam (*) is used to hold the fascia over the round window niche in place. Incudostapedial joint (IS).

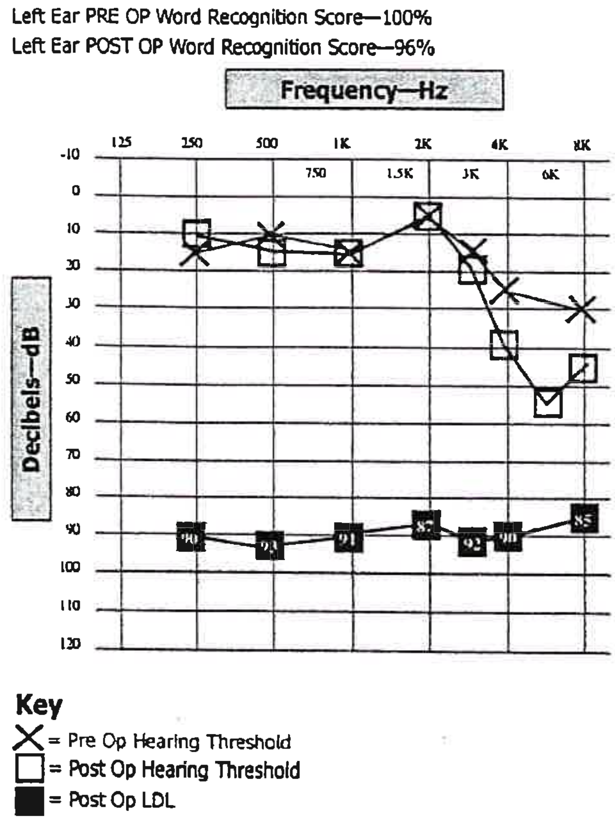

Fig. 4 – Audiometric and loudness discomfort level (LDL) results for Patient 1, left ear.

intolerance and tinnitus after noise exposure from a tank explosion. He had undergone right tympanoplasty for a tympanic membrane perforation a few months after his acoustic trauma. Otoscopic examination was remarkable for a left positive Tullio phenomenon. CT of the temporal bones showed normal superior semicircular canals bilaterally and no other pathologic findings. Pre-operative audiogram showed bilateral mild to moderate high-frequency sensorineural hearing loss with normal tympanograms. Pre-operative vestibular evoked myogenic potential (VEMP) was normal on the left side and absent on the right side, and electronystagmography (ENG) revealed intact and normal bilateral vestibular function. After informed consent was obtained, left round and oval window reinforcement was performed for a suspected perilymph fistula. Post-operatively, hearing remained unchanged except for mild loss in the high frequencies (Fig. 4). The patient did not notice any hearing loss after surgery. Post-operative LDLs were above 90 dB HL in all frequencies except at 2000 and 8000 Hz in the left ear (Fig. 4). Pre-operative LDLs were not routinely obtained at the time of the patient's initial presentation and are not available for comparison. Hyperacusis survey results improved from 33 points pre-operatively to 12 points post-operatively. Post-operatively, the patient no longer exhibited a left positive Tullio phenomenon. He reports continued marked improvement of his hyperacusis and tinnitus, as well as quality of life, at 4 years after surgery.

2.4. *Patient 2*

A 75-year old female presented with a 15-year history of noise intolerance with associated tinnitus that was worse in the right ear. Otoscopic examination was unremarkable bilaterally, but sensitivity and wince aversion to 512 Hz tuning fork testing was noted. CT of the temporal bones showed thinning of the roofs of the superior semicircular canals bilaterally, but no frank dehiscence. There were no other pathologic findings noted on CT imaging. Pre-operative audiogram showed bilateral moderate to moderately severe high-frequency sensorineural hearing loss with normal tympanograms. VEMPs were normal on the left side, while showing reduced thresholds on the right side. After informed consent was obtained, right round and oval window reinforcement was performed. One week after surgery, the patient noted a marked improvement in the ability to tolerate noise in the right ear. Post-operative right LDLs remained under 90 dB HL in all frequencies but improved by an average of 8.67 dB (Fig. 5). As a result of her improved noise tolerance, the patient subsequently elected to have the same procedure in the left ear, which was performed two months later. Post-operative left LDLs improved by an average of 7.14 in the left ear, and improved above 90 dB HL in four frequencies (Fig. 6).

Post-operative audiogram showed no changes in pure-tone thresholds except in the high frequencies in the right ear (Fig. 5). Post-operative audiogram results for the left ear are not yet available. Hyperacusis survey results improved from 29 points pre-operatively to 16 points post-operatively, after both ears were operated on. The patient reports no change in hearing and continued improvement in her hyperacusis and quality of life at 4 months after her first surgery in the right ear, and 2 months after her surgery in the left ear.

Right Ear PRE OP Word Recognition Score—92%
Right Ear POST OP Word Recognition Score—88%

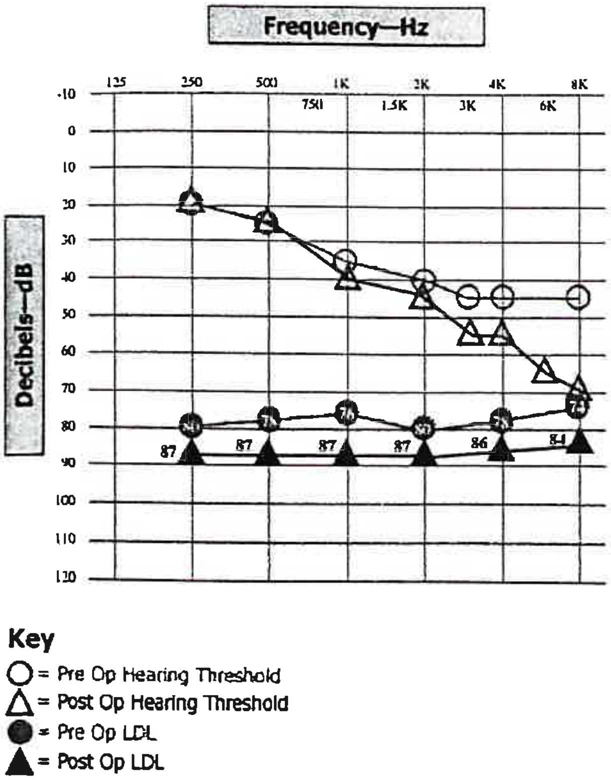

Fig. 5 – Audiometric and loudness discomfort level (LDL) results for Patient 2, right ear.

3. Results

A total of three ears in two patients were operated on by the senior author. Both patients who underwent round window niche and oval window reinforcement with temporalis fascia reported marked improvement in hyperacusis, as well as improved ability to participate in normal daily activities, such as using the telephone and going to restaurants. Both patients reported no negative effects and no subjective changes in their hearing as a result of the procedure, and would recommend the procedure to others suffering from hyperacusis. There were no surgical complications.

4. Discussion

The pathophysiology of hyperacusis remains a heavily researched topic, but is generally accepted to be related to an increase in auditory responsiveness arising from auditory neuronal degeneration. Recent animal studies have shown increased hyper-responsiveness and startle behavior induced by noise exposure causing degeneration of the cochlear nerve [18–20]. Hyperacusis and tinnitus often occur together, and studies have confirmed the involvement of peripheral neural degeneration in the development of both symptoms [8]. Although frequently associated with tinnitus and hearing loss, hyperacusis is also found in individuals with normal hearing. Hyperactivity within the central auditory system has been noted in subjects with clinically normal hearing thresholds [18,21].

Hyperacusis is a common symptom among individuals with SSCD. It is hypothesized that blockage of the round window leads to a reduction in compression-related volume displacement, which causes conductive hyperacusis in patients with SSCD [14]. The success of round window reinforcement in improving the symptoms of SSCD [12,13] led the senior author to consider a similar procedure (round and oval window reinforcement) for patients with hyperacusis without evidence of SSCD.

It is unclear how reinforcement of the round and oval windows leads to an improvement in hyperacusis in patients without SSCD. "Patching" the round and oval windows with temporalis fascia changes the compliance of both windows and renders the inner ear less sensitive to the effects of sudden changes in sound and pressure. Also, it is possible that placement of the temporalis fascia tissue over the round and oval windows acts to dampen the sound waves, which in turn reduces the sensitivity to sounds entering the inner ear fluids. Dampening of the round and oval windows could have decreased the hyper-responsiveness of the central auditory system causing hyperacusis through an unknown mechanism. Audiometric testing indicated a slight decrease in hearing for the high frequencies which was not clinically noticed. Patients stated their hearing was unchanged after the procedure.

Left Ear PRE OP Word Recognition Score— 88%

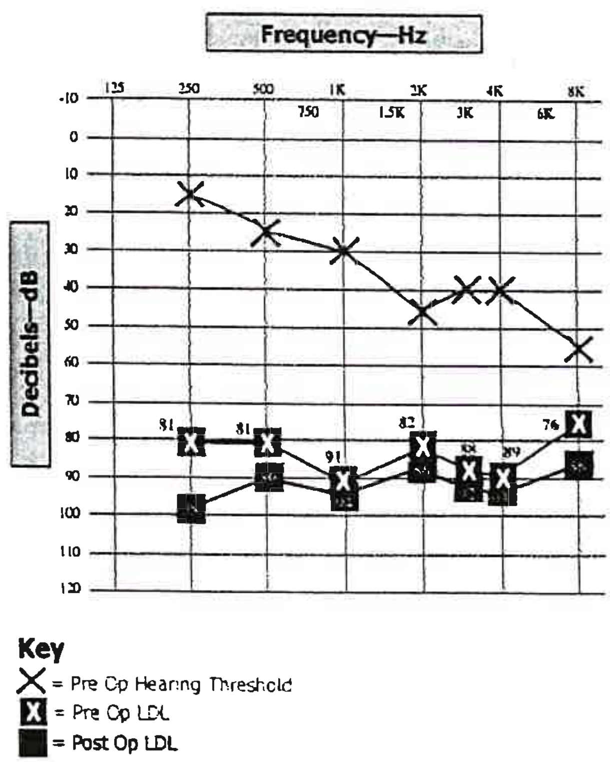

Fig. 6 – Audiometric and loudness discomfort level (LDL) results for Patient 2, left ear.

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Because of the excellent results in our two patients, IRB approval was recently obtained (Sarasota Memorial Hospital) for a prospective study of 20 patients using round window and oval fascia reinforcement to further study the efficacy of this surgical approach in patients with intractable hyperacusis that have failed usual therapy.

5. Conclusion

In conclusion, the authors are the first to report success in improving hyperacusis in two patients without evidence of SSCD using the minimally invasive technique of transcanal round and oval window reinforcement using temporalis fascia. It is hypothesized that round and oval window reinforcement resulted in the reduction of conductive hyperacusis secondary to compression-related volume displacement, or that changing the compliance of the round and oval windows decreases the over-responsiveness of the central auditory system through a mechanism that is not understood. These results are encouraging and suggest that a reliable and minimally invasive surgical quick option exists for patients suffering from hyperacusis after usual forms of long term therapy fail. The long term effects of round and oval window reinforcement for individuals with hyperacusis but without SSCD or other inner ear alterations are unknown at this time. As more subjects with hyperacusis are studied, the efficacy of round and oval window reinforcement in reducing noise intolerance can be further evaluated.

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