

Evaluation and Treatment of Severe Hyperacusis

Michael Valente*
Joel Goebel*
Diane Duddy*
Belinda Sinks*
Judy Peterlein*

Abstract

A 52-year-old male was evaluated by the authors after initially reporting fullness in his left ear while traveling on an airplane. A unique feature of the patient's complaint was the development of severe bilateral hyperacusis (loudness discomfort levels of between 20–34 dB HL) in spite of the fact that the hearing loss was initially reported in the left ear. To achieve loudness comfort, the patient was initially fit with ER-25 musician earplugs that proved to be unsuccessful. The patient next purchased earplugs and earmuffs from a gun shop in order to obtain relief from the pain and discomfort caused by his exposure to everyday environmental sounds. This paper describes the use of hearing devices that proved to be effective in providing attenuation sufficient that the patient rarely needs to rely on earplugs and earmuffs for relief from his hyperacusis.

Key Words: Central hyperacusis, herpes zoster, hyperacusis, loudness discomfort level, musician earplugs, phonophobia, Refuge®-hyperacusis instrument, tinnitus retraining therapy

Abbreviations: CBC = complete blood count, ESR = erythrocyte sedimentation rate, LDL = loudness discomfort level

Hyperacusis (also called “dysacusis,” “phonophobia,” and “hyper-recruitment”) has been reported in patients with acute facial paralysis (Citron and Adour, 1977), Bell's palsy (Gavilan et al, 1988), G_{M1} gangliosidosis type 2 (Gascon et al, 1992), herpes zoster oticus (Byl and Adour, 1976), Ramsay Hunt syndrome (Wayman et al, 1990), lyme disease, endocrine and metabolic disorders, cerebrovascular changes, infectious diseases (Niels et al, 1999), head trauma (Ceranik et al, 1998), idiopathic perilymphatic fistula (Fukaya and Nomura, 1988), acoustic trauma (Axelsson and Hamernik, 1987), Meniere's disease and fibrositis syndrome (Hadj-Djlani and Gerster, 1984), and Williams syndrome (Klein et al, 1990; (Nigam and Samuel, 1994).

Hyperacusis has been defined as a disproportionate growth in subjective loudness of sounds. Simply put, it is a response on the part of an individual that sounds judged “soft” or “comfortable” to a listener with normal hearing are judged “uncomfortable” or “painful” to a patient experiencing hyperacusis. Patients experiencing this problem often avoid social interactions or remove themselves completely from what were once enjoyable situations (e.g., music, theater, restaurants, lectures, etc.). In severe cases, individuals might wear earplugs and/or earmuffs as a way to protect themselves from environmental sounds. In these severe cases, even conversational speech is uncomfortable.

The prevalence and cause(s) of hyperacusis are unknown. There has been speculation, however, that hyperacusis may be related to hypersensitivity of hearing or distortion of the neural coding of the auditory input causing abnormal growth in loudness. There has been a recent trend to suggest that hyperacusis is related to the failure of the central nervous system to habituate the startle response (i.e., “central hyperacusis” [Klein et al, 1990] or “central hyperexcitability” [Phillips and Carr, 1998]).

*Department of Otolaryngology—Head and Neck Surgery, Washington University School of Medicine, St. Louis, Missouri

Reprint requests: Michael Valente, Department of Otolaryngology—Head and Neck Surgery, Washington University School of Medicine, Box 8115, 660 S. Euclid Ave., St. Louis, MO 63110

The term "central hyperacusis" has been narrowed to individuals whose hearing is within normal limits but who experience intolerance to the sounds due to reduction of serotonin (5-HT function) metabolism within the forebrain (Marriage and Barnes, 1995). According to Marriage and Barnes (1995), "subjects complaining of auditory over-sensitivity, who have no other peripheral auditory or vestibular symptoms, should be considered 'central hyperacusis'" (p. 917).

The treatment of hyperacusis is not universally agreed upon. One approach is to simply assure the patient that the presence of hyperacusis is not indicative of any serious underlying disorder. Another approach advocated by Hazell and Sheldrake (1991), Sandlin and Olsson (1999), Byrne and Dirks (1996), Jastreboff et al (1996), and Vernon (1987) is to train the patient to change his/her loudness sensitivity so he/she can gradually tolerate greater sound levels so that normal sound environments are not uncomfortable. In this approach, the patient uses tinnitus maskers to change the loudness sensitivity ("hyperacusis desensitization" or "successive approximation") of the hyperacusis patient. This technique requires long-term exposure to broadband noise, starting at low levels with a gradual increase over time. Hazell and Sheldrake (1991) reported that using this technique improved the tolerance for environmental sounds in 27 of 30 patients. Also, after treatment, the loudness discomfort level (LDL) was increased on average by 5 to 10 dB. Jastreboff et al (1996) reported increasing the LDL, on average, by 14 dB, and, in some cases, the LDL was increased by as much as 30 dB by using similar techniques.

CASE REPORT

Subject

In the latter part of March 1997, this 52-year-old male seminary professor reported fullness and tinnitus in his left ear while traveling on an airplane. A few days later, he noticed decreased hearing in his left ear, nausea, vomiting, distortion of loud sounds, and mild dizziness. This progressed rapidly to greater hearing loss, louder tinnitus, and increased distortion. He denied any relationship of the tinnitus to a Valsalva maneuver he applied during the flight. This is important because the presence of tinnitus following a Valsalva maneuver might have implied a perilymphatic fistula. Initially, he attributed these symptoms to the flu. They continued for about 1 week before he reported these

symptoms to his primary care physician, who then referred the patient to the second author. It should also be noted that the patient reported a bout with shingles (herpes zoster) on his neck and shoulders in November 1996.

PATIENT EVALUATION

Medical Examination

On April 14, 1997, the patient was evaluated by the second author (JG) for an otoneurologic examination. There was no spontaneous gaze, positional, or Hallpike-induced nystagmus, and cerebellar and Romberg's tests were normal. In addition, facial nerve function was normal. At the conclusion of this examination, the physician ordered several blood tests (complete blood count [CBC] with differential and erythrocyte sedimentation rate [ESR]) to rule out infections, inflammation, or other disorders of the blood as possible causes for the patient's symptoms). At this point, the otologist felt that a fistula was unlikely because pneumatic otoscopy was normal and pressure-related symptoms were absent. Early Meniere's disease remained a possibility because of the symptoms of unilateral hearing loss, tinnitus, and vertigo.

Audiometric and Electrophysiologic Evaluations

In addition to the blood tests, a comprehensive audiometric evaluation, auditory brainstem response (ABR), and electrocochleography (ECOG) were ordered. Finally, the patient was placed on diazide and a methylprednisolone dosepak and a second audiometric examination was ordered for the following day (April 15) to determine whether these drugs improved the hearing loss and related symptoms.

The initial audiometric evaluation on April 14, 1997 (Fig. 1) revealed normal hearing in the right ear (not reported in Fig. 1) and a mild (500, 6000–8000 Hz) to moderate (750–4000 Hz) sensorineural hearing loss with a trough configuration in the left ear. The word recognition score for the left ear, using a recorded version of a female talker of the Northwestern University Auditory Test No. 6 word list, was 64 percent presented at most intelligible level. The tympanogram was within normal limits. Contralateral and ipsilateral reflexes were indicative of sensorineural hearing loss of cochlear origin and reflex decay was negative at 500 and 1000 Hz.

A repeat audiogram was performed on April 15, 1997 (see Fig. 1). This revealed improved air-conduction thresholds at 500 and 1000 Hz

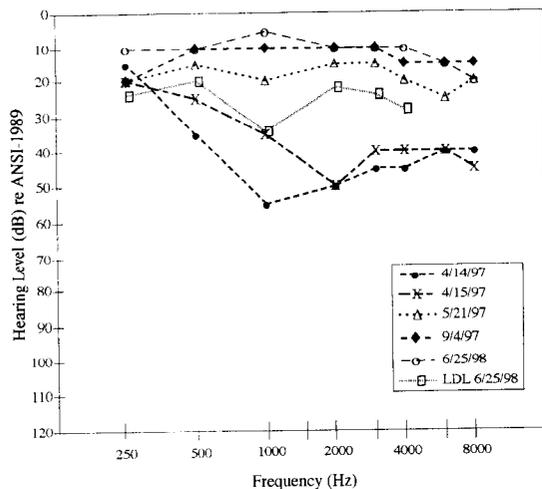


Figure 1 Serial audiograms for the left ear from 4/14/97 to 6/25/98. Also included is the measured LDL for the left and right ears at 250 to 4000 Hz (both ears were identical).

and the word recognition score improved to 84 percent. ABR testing was completed (click stimuli at a presentation level of 70 dB nHL and 80 dB nHL for the right ear and 85 dB nHL and 95 dB nHL for the left ear; stimulus repetition rates were 11.1 and 67.1/sec). Test results were within normal limits for the right ear. Results for the left ear revealed that waves I, II, and III were present at normal absolute latencies with a slight delay in wave IV latency (6.04 msec; normal = 5.70–5.96 msec) and a significant delay in wave V (7.06 msec; normal = 6.35–6.80 msec) latency. The I–III and I–V interpeak latencies were prolonged (I–III was 2.72 msec; normal = 2.18–2.52 msec; III–V was 2.14 msec; normal = 1.80–2.14 msec; I–V was 4.86 msec; normal = 3.98–4.40 msec), and right and left wave V absolute latencies were significantly asymmetric. These results were consistent with left retrocochlear dysfunction. ECOG testing was completed using an ear canal tiptrode referenced to a surface electrode placed at the vertex and contralateral earlobe with the ground electrode place on the forehead. Alternating polarity click stimuli were delivered at presentation levels of 95 and 100 dB nHL and 85 and 90 dB nHL. Summating potential/action potential amplitude ratios were 0.25 and 0.35 for the right and left sides, respectively. These results were within normal limits.

Radiographic Tests

Due to the abnormal ABR in April 1997, an MRI with and without gadolinium was ordered

to rule out a cerebellopontine angle mass or abnormal enhancement of the VIIIth cranial nerve. Results were within normal limits. In November 1997, the patient, for the first time, reported bilateral hyperacusis. To rule out dehiscence superior canal syndrome (an abnormality in the semicircular canal that can make a patient experience dizziness to low-frequency sound), a computed tomography scan of the temporal bones was ordered without contrast. The result was normal.

Blood Tests

In April 1997, at the initial visit, a CBC with differential and ESR tests was ordered to rule out infections, inflammation, or other disorders of the blood as possible causes for the patient's symptoms. The results were normal. In February 1998, the patient reported the recurrence of bilateral hyperacusis and was now using hearing protectors. In order to rule out autoimmune disease of the inner ear, three blood tests (lymphocyte transformation test, antigen 68 kD antibody test, and a repeat ESR) were ordered. All test results were normal.

Follow-Up Audiometric and Electrophysiologic Tests

A follow-up audiogram obtained on September 4, 1997 (see Fig. 1) showed significant improvement in hearing in the left ear. However, the patient reported increased distortion in his left ear and slight distortion in the right ear to virtually all environmental sounds. An ECOG was repeated and results were within normal limits. Additional audiometric evaluations were completed between September 4, 1997 and June 25, 1998 (see Fig. 1). Pure-tone thresholds were within normal limits, bilaterally. During one of these evaluations, acoustic reflex thresholds were attempted but terminated because the patient began to cry due to the pain caused by the stimulus level (70 dB HL). The patient continued to report bilateral distortion and severe hypersensitivity to environmental sounds and conversational speech. At this time, the patient was not using any method to attenuate environmental sounds because (as will be described in a later section) custom-made musician earplugs did not provide sufficient protection. He stated that he was trying to "cope" with the problem.

TREATMENT

Pharmaceutical

After the initial otologic examination, a diuretic (diazide) and methylprednisolone dosepak were prescribed. It was felt by the otologist that one or both of the drugs may have contributed to the return of normal hearing as illustrated in Figure 1 (April 15, 1997).

The continued presence of bilateral hyperacusis in light of an initial unilateral hearing loss remained puzzling. At that time, it was felt that the resulting hyperacusis was probably an expression of central hyperacusis.

Musician Earplugs

For 1 month, the patient followed the regimen of prescribed drugs and completed the various radiographic, audiometric, and electrophysiologic tests. The patient was evaluated in Audiology on May 21, 1997, because he was still very much bothered by the annoyance and pain caused by environmental sounds. At that visit, one of the audiologists (BS) recommended purchase of a ER-25 custom earmold for the left ear. This proved to be unsatisfactory because the patient still experienced sounds that were perceived as painful. On December 4, 1997, an ER-25 custom earmold was fit to the right ear in the hope that using both ER-25s would provide greater benefit. The patient tried these for several months, but, unfortunately, they did not provide attenuation that was satisfactory to the patient.

“Electronic” Attenuator

In June 1998, the patient was referred to the first author (MV) by the second author (JG) to determine if any other treatments were available for the patient. At the time of this visit, the patient was wearing earplugs and earmuffs that he purchased at a local gun shop because all sounds were intolerably loud. It was clear during this visit that even “everyday” sounds were perceived as unbearable (e.g., closing the door of the counseling room, water running in the sink, shoes hitting the linoleum floor) even though he was wearing earplugs and earmuffs. Also, during this conversation, the first author had to whisper the questions because normal, conversational speech caused discomfort even though he was wearing earplugs and earmuffs. The patient reported that because of the hyper-

acusis he could no longer go to the movies, allow his wife to play the piano, go to a restaurant, or attend a lecture. In short, he could no longer enjoy activities that were at one time enjoyable to him and his family. In addition, his self-confidence was affected when he wore the earplugs and earmuffs in public.

At this same visit, LDLs for the loudness judgment of “loud, but okay,” were measured for each ear at 250 to 4000 Hz in 2-dB increments (open squares in Fig. 1). The resulting LDLs were identical for each ear and were between 20 to 34 dB HL. Clearly, these LDLs were extremely low and supported the patient complaint of hyperacusis. In addition, because of the presence of normal hearing, the resulting LDLs supported the probability that this patient had central hyperacusis.

The patient was counseled extensively on a new device that had recently become available for patients with hyperacusis. He was informed that the clinician had no prior experience with this device but was interested in pursuing the potential benefits provided by this technology. The patient was very enthusiastic. From the beginning, the primary goal for both the clinician and patient was for the devices to provide a level of attenuation that would be at least equal to the attenuation currently provided by the earplugs and earmuffs. From the patient’s perspective, achieving this goal would allow him to improve the self-consciousness he felt when people stared at him when wearing the earplugs and earmuffs in public places.

In July 1998, custom-made binaural Micro-Tech® Refuge-hyperacusis instruments were fit. This device is available as a full-concha, unvented in-the-ear instrument provided with a soft flexible canal. The lack of venting and flexible canal assures the user that the only sounds entering the ear canal will arrive via the signal processing of the instrument and not directly into the ear canal from outside. According to the manufacturer, the input/output curve at 2000 Hz provides a 10-dB increase in output with a 40-dB increase in input (i.e., 4:1 compression ratio). For example, if the volume control is at half rotation, an input level of 50 dB SPL would receive just enough amplification to compensate for insertion loss (i.e., attenuation of the input signal caused by inserting the hearing instrument into the ear canal). With the same volume control setting, an input of 70 dB SPL would be attenuated to 62 dB SPL and an input of 90 dB SPL would have an output of 70 dB SPL. The output sound pressure level with a 90-dB input

(ANSI,1996) is between 52 to 67 dB SPL, depending on potentiometer (maximum attenuation with counter-clockwise rotation) and volume control adjustment. Currently, the circuit does not attenuate soft input levels (high-frequency average full-on gain is 1 dB). Finally, the device provides wide dynamic range compression and variable release time (attack time = 150–985 msec; release time = 1900–2000 msec).

The devices were fit and the patient was counseled on their use and care. Because of the patient's past reactions to stimuli during formal testing (i.e., crying during reflex testing), it was decided by the patient not to objectively evaluate the performance of these devices (i.e., LDLs measured for frequency-specific and broadband signals for unaided, earplug + earmuff and Refuge[®]-hyperacusic instruments). Rather, the patient would subjectively critique the devices for the next 2½ months (he was leaving for a trip to England the next day). In September 1998, the patient returned from England and said he was very pleased with the performance of the devices and decided to purchase them. He reported that the instruments provided a level of protection that was at least as good as he achieved with the plugs and muffs.

Update

The patient returned in September 1999 for a follow-up visit. He reported that he wears his binaural Micro-Tech[®] Refuge-hyperacusic hearing instruments all of the time and finds them very helpful. Occasionally, he still uses his circumaural headset in combination with his instruments. He still cannot listen to music, attend concerts, or go to restaurants having music in the background, movie theaters (because of music in the background), or graduation ceremonies. While eating, he cannot hear anyone talking because of the occlusion effect created by wearing the hearing instruments. He wears one of the hearing instruments (right ear because he typically sleeps on his left side) while sleeping so that traffic noise outside his house does not disturb him.

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