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20 Questions

One of the goals of Page Ten is to keep you informed of the latest testing and treatments for auditory disorders. One intriguing area that has emerged in recent years is the diagnosis and management of the patient with hyperacusis. Both the condition and the patient with it are often misunderstood.

The term is derived from Greek. "Hyper" is a prefix signifying above, beyond, or excessive. "Acusis" is sense of hearing. Logically then, we would think of the hyperacusic patient as someone with extra-good hearing (a built-in Whisper 2000). In fact, years ago I recall that when a child would show up in the clinic with a consult saying "rule out hyperacusis," we would test the child's hearing, observe thresholds in the 5-dB to 10-dB range, and tell Mom and Dad that all was well. But all may not have been well.

Since then, we have learned that what troubles the hyperacusic patient most is not "super sensitivity" for soft sounds, but rather the increased loudness perception for average or slightly loud sounds. In other words, sounds that you or I may find comfortable could be uncomfortably loud to the hyperacusic patient.

As you can imagine, treatment of the hyperacusic patient is a challenge. We've found an individual, however, who not only works with these patients on a regular basis, but is willing to tell his story on Page Ten. **James W. Hall III**, PhD is

no stranger to *The Hearing Journal* (although he may appear to be in his new photo!) Dr. Hall is Associate Professor in the Division of Hearing and Speech Sciences at Vanderbilt University and in the Department of Otolaryngology at the Vanderbilt School of Medicine as well as being director of the Vanderbilt Balance and Hearing Center.

If you don't know Jay from his successful books, you've probably been to one of his seminars enrolled in one of his popular hands-on workshops in "Music City." He tells us that his interest in the hyperacusic patient was an outgrowth of the work he has been doing with CAPD and tinnitus. As you read Jay's article, you'll see there is some overlap among these auditory conditions.

Hyperacusis is real, and there are probably more people suffering from this disorder than any of us knows. I hope this excellent Page Ten article will increase your awareness of the problem and provide you with some diagnostic and treatment strategies.

Gus Mueller, Editor
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Hyperacusis...it's real and it can hurt!

By James W. Hall III

1 What is hyperacusis? Is it increased sensitivity to faint sounds?

No, hyperacusis is not hypersensitive, or extra-good, hearing. Hyperacusis is reduced tolerance to suprathreshold sounds. For the hyperacusic patient, moderate- to high-intensity sounds, which would be easily tolerated by other persons, are bothersome, annoying, uncomfortable, or even painful.

2 What causes hyperacusis?

The origin of hyperacusis is in the central auditory nervous system, rather than in the ear (the cochlea). Certain everyday sounds of moderate intensity are perceived by the patient as too loud. One way of describing this problem is to state that the central auditory system gain or amplification for sound stimulation is excessively high.

3 If hyperacusis has a basis in the central auditory nervous system, is it associated with any other CNS disorders?

Yes. There are a variety of neurologic conditions associated with hyperacusis, among them post-traumatic stress disorder, chronic fatigue syndrome, Tay-Sach's disease, some forms of epilepsy, benzodiazepam dependence (Valium is diazepam), depression, and migraine headaches (for review see Marriage & Barnes[1]).

A disruption or disturbance in a neurotransmitter—5-hydroxytryptamine (5-HT) or serotonin—is a common neurochemical feature of each of these disorders. In some way, this neurotransmitter influences a person's sensitivity or reaction to sensory stimuli. The exact CNS physiologic mechanism underlying hyperacusis, however, is not known at this time.

4 How does hyperacusis differ from loudness recruitment?

Patients with hyperacusis typically have normal hearing, whereas loudness recruitment is associated with sensory hearing loss and, in particular, outer hair cell damage and dysfunction. A better term for loudness recruitment is "abnormal growth of loudness." People with hyperacusis also have an abnormal loudness growth function. However, it is a very different pattern from that observed with the typical person with cochlear pathology.

There are several peripheral disorders that may also have, as a component, increased sensitivity to sounds, even though hearing sensitivity is normal. Examples include patients with facial nerve paralysis (and absence of the stapedial acoustic reflex), as in Bell's palsy, and status post-stapedectomy, after the stapedial tendon has been severed surgically.[2–4] These are not the kinds of hyperacusis patients who present the real challenge to audiologists.

5 What audiometric test protocol do you use in the assessment of hyperacusic patients?

A very comprehensive basic audiologic assessment is essential for starters.[5,6] The test battery should include pure-tone audiometry and otoacoustic emissions (transient or distortion product) to rule out sensory hearing impairment, some measure of word recognition, and also tympanometry to assess middle ear status. *As a rule, acoustic reflexes should not be recorded, since the required intensity levels (80 dB and greater) may cause the hyperacusic patient undue discomfort, distress, or pain.* Loudness discomfort levels (LDLs) for pure-tone signals of 1000 Hz and higher are often very helpful in documenting decreased tolerance to sound. Use of an ascending approach for this procedure is, of course, recommended.

6 Is there an audiometric pattern that is characteristic of hyperacusis?

Although there is no single distinct collection of findings invariably associated with hyperacusis, most patients have perfectly normal hearing sensitivity, normal or even larger than normal OAE amplitudes at some test frequencies, and unusually low LDLs.

A pattern of audiometric findings is shown in Figure 1 for one of my pediatric hyperacusic patients. He is a 10-year-old boy who 6 months earlier had suffered a severe head injury in a motor vehicle accident and was referred to me for assessment of possible central auditory processing disorder (CAPD). History included intolerance to everyday sounds, such as vacuum cleaners, water running from the faucet, and telephones ringing. The patient refused to go to the school cafeteria or to attend athletic events in the school gym because he was fearful of loud sounds.

Note that this child's hearing sensitivity was within normal limits and, in fact, slightly better than expected in the high frequencies. Middle ear function was normal. However, LDLs were unusually low for both ears. Also, distortion product OAE amplitudes for high-frequency stimuli were well above the upper limit of the normal region.

7 What kinds of sounds bother a person with hyperacusis?

Some of the same sounds mentioned earlier for the 10-year-old boy. I've had patients who were excessively bothered, even terrified, by dog barks or laughter. Sounds that are ultra-high-pitched seem to be particularly bothersome, such as signals emitted by televisions, security systems, or answering machines. The term "shrill" is often used by patients in describing extremely annoying or even painful, sounds. One of my pediatric patients couldn't stand the "popping" sound of a cooling car engine in the garage. Remember, their LDLs may be in the 60 dB (HL) to 70 dB (HL) range; many environmental sounds exceed this level.

8 These complaints sound like "phonophobia." What is the difference between hyperacusis and phonophobia?

Phonophobia and hyperacusis are sometimes used interchangeably,[1] whereas some authors make a distinction between the terms.[7] Hyperacusis might be thought of as an *intolerance* to most moderate- to high-intensity sounds, while phonophobia is a *fear* of certain sounds, even at relatively modest intensity levels.

According to Jastreboff, phonophobia involves activation of the autonomic and limbic regions of the nervous system.[7] These regions of the brain mediate emotional and/or fearful reactions to sound. It's not uncommon to hear patients describe their discomfort with a variety of everyday sounds, and fear of some, and yet then express pleasure at listening to their screaming and shouting grandchildren or in cranking up their favorite music on the stereo. As an aside, *photo*phobia (hypersensitivity to light) may be associated with hyperacusis and/or phonophobia.

9 Is there any connection between hyperacusis and tinnitus?

Yes. Hyperacusis and tinnitus often coexist. In fact, a patient's complaint of hyperacusis (without tinnitus) should be taken very seriously, in part because hyperacusis may be a precursor of tinnitus. "Increased central gain" is probably a factor in hyperacusis and in some patients with very bothersome tinnitus.[8, 9]

10 Is hyperacusis mostly an adult problem, or can children also be hyperacusic?

Adults may be more willing and able to express their concerns about hyperacusis, but the problem can certainly affect children as well. Parents of hyperacusic children may describe them as "auditorially defensive." Hyperacusic children may cover their ears with their hands or run out of the room if there is an annoying sound, rather than calmly and rationally describing their intolerance to the sound.

One of my 9-year-old patients couldn't tolerate a sound in a film shown in his classroom. According to his mother's emotional report, "He covered his ears and ran out of the classroom, down the hallway, out the front door of the school, and part-way across a busy four-lane road!" Naturally, I'm taking this child's hyperacusis very seriously.

11 Is hyperacusis more common in certain pediatric audiology populations?

In my experience, hyperacusis seems to occur more frequently among children who are referred for CAPD assessment, who are also in the differential diagnosis for learning disability and attention deficit disorder (all these disorders involve the central nervous system), in children with clear neurologic insult (like the head-injured boy described earlier), and in children with a history of chronic middle ear disease and conductive hearing loss, with recent resolution to normal hearing sensitivity. For this last group, chronically reduced auditory input may have led to increased central gain. Hyperacusis is also a characteristic feature or symptom of one pediatric disorder—Williams Syndrome.[1, 10]

12 I'm not familiar with Williams Syndrome. Would you describe it briefly?

I'm glad you asked. Williams Syndrome, first described in 1961, is characterized by decreased growth, prominent lips and atypical facial appearance, a hoarse voice (especially in girls), and other significant anomalies, among them hypercalcemia, renal hypertension, bladder diverticula, and heart defects (subaortic stenosis). The patient usually has some degree of mental retardation.

The incidence of Williams Syndrome is estimated at between 1:20,000 and 1:50,000, although the syndrome is probably underdiagnosed. Life expectancy is normal unless the patient has a serious heart defect.

For more information on this interesting syndrome, check the Internet or contact: Williams Syndrome Foundation, University of California, Irvine, CA 92697-2310 (Tel: 714-UCI-7259; Fax: 714-824-7259).

13 What about autistic children and those with pervasive developmental delay? Do they also experience hyperacusis or phonophobia?

Clinical experience strongly suggests the existence of hyperacusis, phonophobia, or some form of hypersensitivity to sound in some children with autism or autistic-like behaviors. Auditory integration therapy (AIT) is, in part, performed to desensitize such children to sound, presumably by manipulating "central gain."

14 Would (AIT) be an appropriate treatment approach for children with hyperacusis?

Many audiologists have fundamental methodologic questions and concerns about AIT. AIT is viewed as experimental and is neither recommended nor approved by professional audiology organizations, pending publication of findings for controlled clinical studies documenting its efficacy.

15 As an audiologist, what *can* I offer a patient with hyperacusis?

First and foremost, offer *understanding*. Hyperacusis is real for the patient, and you can take a big therapeutic step forward by acknowledging the seriousness and validity of his/her problem.

Also, take a very thorough medical, audiologic, and, to a lesser extent, psychological *history*. Seek out any reference to diseases or disorders, or medications, that might be associated with hyperacusis. And, specifically ask about tinnitus.

As I've already mentioned, you should perform a comprehensive *audiologic assessment* to rule out cochlear or retrocochlear dysfunction. Don't forget to include OAE recordings and frequency-specific LDL measurements in your test battery. Of course, make a medical, possibly otologic or even psychiatric, referral if it's indicated by the history or audiologic findings.

16 What do you do with all of the information from the history and the hearing assessment?

Counsel the patient. Take some time to explain the findings and as much as you can about their auditory system. Use your handy AAA ear chart, and any other visual aids that might get the points across. Provide good and accurate advice and information, both orally and in the form of a brochure or hand-out.

17 Since hyperacusic patients are bothered by loud sounds, wouldn't hearing protection be part of their management?

Quite the contrary. In your counseling, unequivocally advise the patient to *avoid silence*. The patient's natural tendency is to seek out quiet. Some patients wear earplugs almost constantly to avoid as much exposure to sound as possible. They might even show up for their audiology appointment wearing earplugs and/or earmuffs. This strategy is, in fact, counterproductive, because it is likely to prompt the central nervous system to increase gain even further.

The patient should consider hearing protection only when there is the likelihood of exposure to high-intensity sounds that could potentially cause noise-induced hearing loss or during extended periods of bothersome noise (e.g., riding in an airplane or car).

18 How can a patient avoid being in quiet places sometimes?

The patient should strive to always be around low-level and pleasant sound. At home, or in a quiet workplace, the patient can play a radio at a low volume. The use of environmental noise generators (available from many stores and mail-order companies) should be strongly encouraged, especially at night in the bedroom when the patient may be "exposed" to silence. Finally, it is quite appropriate, and often very effective, to fit the patient with *low-level noise generators*, after all the above steps have been completed.

19 Tell me more about these low-level noise generators. Do they offer a "cure" for hyperacusis?

There is no overnight cure or treatment for hyperacusis, or tinnitus for that matter. However, constant stimulation over the course of months with very consistent, pleasant, low-levels of noise (essentially white noise) can help the brain readjust to everyday sounds.[6, 9] Documentation of the patient's decreased hyperacusis over time can even be shown in more normal LDLs.

It is very important to stress that the audiologic assessment and extended counseling should always be completed before the patient is fitted with noise generators.

Several companies market BTE or ITE versions of noise generators (e.g., Viennatone, Starkey, General Hearing Instruments). It's important to verify that the noise intensity level can be increased from 0 dB and that the noise level remains constant at a given VC setting.

20 Any final comments on the audiologist and hyperacusis?

Patients with hyperacusis (hyperacusis plus tinnitus) are a large and, at this time, underserved, patient population. An audiologist providing appropriate services to the hyperacusic patient can strengthen his/her position as an effective entry point to hearing care and can make a real difference in the patient's quality of life.

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