
DISORDERS

HYPERACUSIS

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Auditory Hyperacusis

By Eric Bostwick, AuD

WHAT IS HYPERACUSIS?

Several interpretations and definitions of hyperacusis exist, but there is no definition of the condition that is universally agreed upon. However, hyperacusis is usually characterized as general intolerance to noise and ordinary sounds that triggers a physical and/or psychological response. Broken down: hyper- meaning excessive, and -acusis, referring to sound perception.

HYPER = EXCESSIVE

**ACUSIS = SOUND
PERCEPTION**

Tyler et al. (2014) have suggested that hyperacusis can be divided into four distinct subgroups, and that these can occur individually or in combination:

1. Loudness hyperacusis—sounds are judged by the listener to be uncomfortably loud in comparison to what a person with “normal” hearing would perceive. Individuals with loudness hyperacusis may have hearing loss, but this condition can also occur in individuals that have clinically normal hearing sensitivity.
2. Annoyance hyperacusis—a negative emotional reaction to loud sounds that can manifest as increased irritation, anxiety, or tension.
3. Fear hyperacusis—an aversive response to sounds that results in anticipation and avoidance behavior where the listener takes steps to keep away from certain sounds.
4. Pain hyperacusis—in which the listener experiences pain well below the normal threshold of painful auditory stimuli (approximately 120 dB, or the sound of a jet plane during take-off). What would be described as a “tickle” [37, 42] or discomfort [37, 44] in normal listeners is characterized as a sharp stabbing sensation.

Hyperacusis can be debilitating, and those who have this invisible condition often suffer from a lack of understanding from others when explaining how they feel [34]. Hyperacusis can have a negative impact on overall quality of life, and it can lead to changes in behavior, avoidance of loud situations such as crowds or public transportation, and minimization of interactions with friends, family, and coworkers [36]. If left unaddressed, these adverse effects can eventually lead to diminished socialization, lessened physical activity, poor performance at work and school, and in some cases, financial stress [34]. Furthermore, the condition impacts everyone a little differently, each person’s adaptive strategies can vary



widely, and the disorder can mimic other conditions of the auditory system.

Hyperacusis almost exclusively affects both ears [4], and generally presents as a chronic condition [33]. This important distinction of hyperacusis as a bilateral, persistent condition helps to separate it from other possible differential diagnoses. Hyperacusis is different from phonophobia (an episodic intolerance of sound during migraine attacks which goes away as the attack stops) [6] as well as misophonia (an adverse reaction toward specific human sounds such as eating, breathing or chewing) [24, 26]. It is also distinct from autophony—the temporary perception of one’s voice as hollow, unusually loud, or reverberant. This perception can be due to Eustachian tube dysfunction. The Eustachian tube is a channel that connects the middle ear space to the back of the throat, which normally opens and closes in response to changes in pressure (e.g. when your ears “pop” while on a plane). If this system remains open when it is not supposed to be, sounds can vibrate through the mouth and travel directly to the eardrum [30]. Additionally, autophony can be the result of another group of rare medical conditions called inner ear “3rd window pathologies.” This is sometimes called “conductive hyperacusis.” The scope of this article will exclude discussion on 3rd window pathologies of the inner ear and their effect on the vestibular system, namely superior semicircular canal dehiscence. For more information on this topic and its contribution to dizziness symptoms, please reference Dr. Minor & Dr. Carey’s article on the subject matter here.

The common complaint for each of the above conditions as well as hyperacusis is a report of sounds being loud. Without a clear accepted definition and variable symptomatology, hyperacusis represents a challenging problem in the healthcare community [38]. Hyperacusis patients are commonly seen by a variety of healthcare disciplines, and it can sometimes take several appointments with different providers and specialists to diagnose and treat hyperacusis effectively.

WHO IS AFFECTED BY HYPERACUSIS?

There are few studies that have investigated how widespread hyperacusis exactly is. Part of this lack of understanding is due to the presence of other symptoms that often make the diagnosis difficult. These include: headache, tinnitus (ringing, buzzing, chirping, etc., in the ear), depression [41], migraine [32], stress, anxiety [38], and hearing loss [40] some



of which are not fully understood in their own right. Even with conservative estimates, hyperacusis affects millions of people worldwide. Prevalence estimations range between 2% [35] to 15.2% [14], but it is likely widely misdiagnosed due to the variability in how the condition is reported. There is also variability in how hyperacusis affects the auditory system itself. Studies on the prevalence of tinnitus presenting in combination with hyperacusis range from 40% [2] to 86% [20]. With hyperacusis, tinnitus and hearing loss may also occur in combination, and the relationship between these is still complex and unclear.

Other contributing factors include acoustic trauma such as gunfire and explosions, as well as head trauma, aging [23, 25], and concussions [3]. There may also be a relationship between the disorder and personality traits [41], as well as other medically idiopathic and unexplainable symptoms such as chronic fatigue, and environmental reactions including: chemical sensitivity, “non-specific building-related symptoms,” and exposure to electromagnetic fields [7, 8, 29, 31]. More research into these topics is needed to further characterize hyperacusis and its relation to other medical conditions.

WHAT CAUSES HYPERACUSIS?

The cause of hyperacusis is still a heavily researched topic [38]. Based on available findings, it is likely that hyperacusis has both peripheral auditory system and brain-level contributions. Peripheral theories identify two possible contributing factors: 1) damaged outer hair cells in the inner ear that are responsible for loudness growth and compression lose function, and 2) auditory nerve degeneration distorts the integrity



of the incoming sound as it is being sent up to the brain ^[40]. There is a growing consensus among many audiologists, otolaryngologists, and neuroscientists that hyperacusis is primarily a brain-level disorder. Several studies suggest that hyperacusis is a result of an increase in central auditory “gain,” wherein signals from structures within the brain are increased ^[18].

With respect to pain and hyperacusis, the origin of these sensations is still not well understood. The sensation of pain in the body arises in part from the stimulation of nociceptors, sensory cell endings responsible for transmitting information that can be potentially threatening or damaging. However, none of these receptors are present in the cochlea, the hearing organ in the inner ear ^[40]. Some research has suggested that nerves innervating the outer hair cells can create the sensation of pain for loud sounds by detecting tissue damage ^[15], but more definitive research is needed.

MEDICAL EXAMINATION & QUANTIFYING HYPERACUSIS

Despite the conflicting reports of hyperacusis, formal medical evaluation by a physician should not be discounted. Medical evaluation for patients presenting with hyperacusis symptoms is aimed at delineating hyperacusis from other underlying pathological conditions, many of which can be treated. Physical examination should include looking in the ears, measurement of eardrum mobility, and assessment for vertigo. Medical consultation is also important to rule-out other possible diseases that may be life-threatening. Your doctor may order blood tests, or medical imaging such as a CT scan ^[28], MRI, or MRA ^[34]. You may need a referral to a dentist if temporomandibular joint disorder (TMJ) is suspected ^[10, 19].

Other medical conditions associated with hyperacusis that may require specific testing include: head trauma, depression, PTSD, genetic syndromes, fibromyalgia, chronic fatigue syndrome, generalized anxiety disorder, exhaustion syndrome, irritable bowel syndrome, and multiple sclerosis. Other studies have explored the relationship between hyperacusis and other less understood disorders such as migraines, Meniere’s disease, and phantom limb pain ^[40]. Many of these have been known to cause sensations of dizziness. The challenge that remains when considering all of these underlying disorders is how to quantify something that is seemingly invisible.

Two main approaches are used to quantify loudness hyperacusis in comparison to normal

listeners. These include measuring Uncomfortable Loudness Levels (ULLs or UCLs) to pure tones, as well as complete step-wise assessment of loudness perception as sound intensity increases, called a loudness growth function [34]. It is worth noting that many of these measures in the test booth that use tones do not always translate well to the experience of real-world sounds ^[9, 27, 43]. It is known that there are many individual factors and circumstances that can affect our reaction to a specific sound or class of sounds (e.g. fatigue, tiredness, etc.) ^[40], annoyance can be unrelated to loudness ^[2, 11, 16] and that in many cases our perceived control of the situation is important when judging how annoying sound is (e.g. someone else’s baby crying in an airplane vs. your headphone volume being turned too high—something easily adjustable) ^[11, 13, 17]. There are also many self-report questionnaires available that clinicians can use as tools to help get an idea of how impactful hyperacusis symptoms are on your day-to-day life. These measures can be helpful in quantifying degree of debilitation, and serve as a baseline measure for comparison when measuring treatment effectiveness.

TREATMENT OPTIONS

Hyperacusis is frustrating, but the good news is that there are a variety of treatments that have shown success. Current auditory treatment options for hyperacusis patients include: tinnitus retraining therapy (if tinnitus is present), hearing amplification devices, and gradual sound exposure using noise generators ^[1, 22]. Sound therapy can

TREATMENT OPTIONS

- Tinnitus retraining therapy
- Hearing amplification devices
- Gradual sound exposure
- Sound therapy
- Cognitive Behavioral Therapy
- Social Support
- Medications
- Surgery



include desensitization, or presenting noise at a level that is not challenging to the patient and then slowly raising the volume over time, as well as using steady, comfortable sound levels to help recalibrate the auditory system [4, 21]. Cognitive behavioral therapy (CBT) has also proved beneficial. CBT includes working with a counselor to promote education, develop applied relaxation techniques, progress through exposure to aversive stimuli, and reduce stress. These techniques help to retrain the brain's response patterns to these types of sounds. Social support is also an important part of the rehabilitation process, and can help with getting used to new sounds as they increase in intensity [34].

A common self-treatment of hyperacusis is the use of hearing protection; however, this approach is usually not part of a long-term solution. Instead of addressing the underlying issue, routinely wearing hearing protection can strengthen the reliance on earplugs and earmuffs to function normally in society. Some studies have suggested that they strengthen maladaptive response patterns and reinforce association between the sound itself and distress. Reliance on hearing protection has also been shown to further increase the central gain applied to sounds in the brain and make hyperacusis worse [12, 18, 39]. Various drug therapies are also currently being explored with some promising findings, but more controlled research is needed [34]. Lastly, there are also some reports of success using proactive surgical techniques used for superior semicircular canal dehiscence to help dampen sound waves as they enter the inner ear [38]. Each of these treatments as well as surgical candidacy should be something that is discussed further with your doctor.

CONCLUSIONS & FUTURE DIRECTIONS

Hyperacusis can be discouraging. It can alter the course of one's life and daily routine. Various treatment options are available that span the scope of several medical professions. If you are someone that suffers from auditory hyperacusis, it is always important to try and do the little things on a daily basis to promote general health and wellness—making sure you're reducing stress when possible, sleeping well, eating a healthy balanced diet, staying on top of any other medical conditions you may have, asking questions about any medications you're unsure of, etc. The human body is a complex integrated set of systems, and taking control of these little choices that we all have available to us on a day-to-day basis can help significantly with symptoms and treatment plans.

Future research should aim to better define

hyperacusis and come to a universal consensus on what to include in its symptomatology. This will reduce the number of misdiagnoses and help these patients seek proper treatment. Additionally, more research on both human sound processing and animal models is needed, as there are several aspects and mechanisms of loudness perception that to this day are still not well understood. The good news is that research in this area is growing, and that more and more laboratories and clinics have devoted resources to further investigate hyperacusis over recent years [5]. To further assess these symptoms, full examination by a Neurotologist and Audiologist is recommended as a starting point. If you or someone you know suffers from hyperacusis, sound sensitivity, or sound-induced dizziness, it is recommended that you schedule an appointment with a doctor and discuss these sensations further. You may find an answer to some of the concerns and questions that you are looking for.

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