HYPERACUSIS

What is hyperacusis?

Hyperacusis is a disorder in loudness perception. It is also called decreased sound tolerance or sound sensitivity. People with hyperacusis perceive certain everyday sounds as annoying, uncomfortable, painful, distressing and even terrifying. Hyperacusis can have a significant negative impact on activities of daily living. It can start gradually, or the onset can be sudden.

Trigger sounds are often high-pitched or start abruptly (impulse sounds) such as reversal beeps on trucks, pots and pans clattering, jingling coins, and dogs barking. Some people are bothered by continuous, low-pitched sounds such as computer fans, running water or refrigerator compressors. People with hyperacusis may have an increased awareness of sounds such as the ticking of a clock. Unexpected sounds, such as a balloon popping or screeching brakes, may bring about a strong startle response with hyperacusis.

As well as the more common loudness hyperacusis, a smaller number of people experience pain hyperacusis. With pain hyperacusis, some sounds bring on piercing head pain or migraine.

Some sounds make affected people anxious or fearful. This is called fear hyperacusis (phonophobia). Children with hyperacusis may struggle with sounds such as the washing machine or a hand dryer and refuse to go near the sound source.

Some trigger sounds, even soft ones such as other people chewing or a keyboard tapping, bring on an immediate strong reaction in people affected. This rare condition is referred to annoyance hyperacusis (misophonia). The reaction may range from annoyance to disgust, rage, panic or an immediate urge to flee. Annoyance hyperacusis sometimes has visual or olfactory (sense of smell) triggers.

Some sound-related symptoms may be confused with hyperacusis. These include:

- **Tullio phenomenon** – sound-induced dizziness. It is a rare symptom associated mainly with semicircular canal dehiscence syndrome (SCD).
- **Recruitment** – abnormal increase in the perception of loudness accompanied with hearing loss. With recruitment, loud noises seem uncomfortably loud; however, with hyperacusis, all sounds seem too loud.

Almost everyone finds some sounds annoying. For many, trigger sounds may be the screech of fingernails dragging down a blackboard or the repetitive drip of a leaky faucet. These are normal variations of sound intolerance, not hyperacusis.

Hyperacusis affects people of all ages. It is estimated that about 2 to 3% of people have some degree of hyperacusis. Only a small number of these are severely affected. About half of people with hyperacusis also have hearing loss.

More than 80% of patients with hyperacusis also have tinnitus (ringing in the ears). Hyperacusis can develop before, at the same time as, or after the onset of tinnitus. The frequent combination of hyperacusis and tinnitus suggests they may be caused by a similar mechanism.

What causes hyperacusis?

The mechanisms that provoke sound sensitivity are unclear. One theory is of abnormally increased sensitivity within the brain’s hearing system (increased auditory gain). If the brain were imagined as having a volume control dial, with hyperacusis the dial would be turned all the way up. Sounds that normally would be tolerated are perceived as too loud.

Other theories for why hyperacusis occurs include:

- malfunction of the ear’s protective hearing mechanisms
• damage to a portion of the auditory nerve
• a malfunction of the facial nerve

Decreased sound tolerance can be associated with a number of conditions, some of which are treatable, including:

• **Ear-related (otologic):** Ramsay Hunt Syndrome, Ménière's disease, perilymph fistula, semicircular canal dehiscence (SCD), acoustic trauma (often caused by exposure to high-decibel noise), barotrauma (injuries caused by increased air or water pressure), noise-induced hearing loss, stapedectomy (surgery to remove a small bone from the middle ear to improve hearing), and tympanoplasty (surgery to repair a perforated eardrum).

• **Neurologic:** autism spectrum disorder (ASD), Bell's palsy, carotid or middle cerebral aneurysm, migrainous cerebral infarction, head injury including concussion, Chiari malformation, sympathetic reflex dystrophy, multiple sclerosis, migraine, epilepsy, myasthenia gravis, cerebrospinal hypertonia, primary thalamo deficiency, attention-deficit disorder (ADD), anxiety and depression disorders, post-traumatic stress disorder (PTSD), and complication of spinal anaesthesia.

• **Endocrine:** Addison's disease, panhypopituitarism and hyperthyroidism.

• **Infection:** neurosyphilis, Lyme disease and typhoid fever.

• **Medication:** benzodiazepine and antidepressant withdrawal and acute phenytoin intoxication.

• **Vitamin and mineral deficiency:** magnesium and pyridoxine (vitamin B6).

• **Genetic or congenital:** Williams syndrome, idiopathic hypercalcaemia (Fanconi and Williams-Beuren syndrome), Cri-du-Chat syndrome, Tay-Sachs disease, Cogan syndrome, GM1 gangliosidosis and spina bifida.

• **Other:** temporomandibular joint (TMJ) disorder and fibromyalgia.

The noise of an MRI scan may trigger or exacerbate hyperacusis. Scanning sequences for the internal auditory canal (IAC) are especially noisy. Hearing protection should be available, and equipment can be set up to reduce noise exposure. In some cases, hyperacusis appears without connection to any event or condition.

**Diagnosis of hyperacusis**
Hyperacusis can be assessed by pure-tone audiometry (a hearing test used to identify hearing thresholds), measurement of uncomfortable loudness levels, and by the doctor or audiologist talking with the patient about how they perceive sound.

**Impact of living with hyperacusis**
Some people feel they are sound sensitive but that it has minimal effect on activities of daily living. Those who are more severely impacted may become so fearful of sounds that they withdraw from normal activities in order to avoid sound. Those affected by hyperacusis may report:

- difficulty concentrating
- anxiety or even fear
- tension or even anger

**Treatment and management**
There is no quick cure to eliminate hyperacusis. However, a combination of therapies can improve quality of life by reducing fear and anxiety, developing strategies to cope with uncomfortable sounds and even decreasing sound sensitivity. Judicious use of hearing protection to muffle sounds in loud environments can help. Overuse of hearing protection in quiet environments, however, can trigger hyperacusis and likely makes existing hyperacusis worse.

Most researchers agree that people with tinnitus overuse hearing protection. Ask a hearing specialist for advice on when and how to best use hearing protection.

**Treatment of associated medical conditions**
Treatment options include: counselling, including cognitive behavioural therapy (CBT), to address emotions related to living with hyperacusis as well as any anxiety or depression that may contribute to sound intolerance. CBT can help patients change their pattern of thinking or behavior to lessen the impact of hyperacusis on their life. Note: it can be challenging to find a CBT practitioner who is knowledgeable about hearing disorders.
Education to understand that sound perceived as loud or uncomfortable isn’t necessarily causing permanent damage to hearing. If other people aren’t covering their ears or looking uncomfortable, then the sound likely isn’t doing any damage.

**Sound therapy** to retrain the brain’s auditory processing centres to accept everyday sounds as normal again.

**Sound generators worn in the ear** may help some patients to gradually increase sound tolerance and reduce the contrast between bothersome noise and background noise.

**Modified Tinnitus Retraining Therapy (TRT)** has been shown to have an 85% success rate for people with misophonia. Gradual exposure to increased sound in safe and predictable environments. This may mean slowly increasing sound levels when at home or venturing out with ear protection that can be put on if needed. Good quality electronic earmuffs may be suitable for hyperacusis and tinnitus patients exposed to intermittent noise. Their use can decrease overuse of hearing protection.

Patients who have sound-induced pain and temporary aural fullness (feeling of pressure or fullness in the ear) may be helped by a physiotherapist with special training in temporomandibular joint disorders (TMJ) and cervical (neck) treatment. This is particularly useful for those with a history of head or neck injury. Serious medical problems contributing to ear pain and aural fullness should be ruled out by medical assessment before seeing a physiotherapist for hyperacusis.

What to expect in the future

The speed and degree of recovery from hyperacusis varies from person to person. It may happen over weeks or months. Hyperacusis may improve by giving the ears time out or it may need specific treatment. It may fully resolve or there may be ongoing sound sensitivities necessitating changes in lifestyle or work.

More resources and sources

View more hyperacusis resources as well as sources used for this handout: [https://bit.ly/2QnXgOE](https://bit.ly/2QnXgOE)

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