Hyperacusis and its Management

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Hyperacusis and its Management

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Presentation Outlines

• Definition of Hyperacusis
• Neurophysiologic, peripheral and central causes of Hyperacusis
• Pathological Conditions Associated with Hyperacusis
• Management of *Decreased Sound Tolerance Disorders*
Hyperacusis

• An increased sensitivity to sound
• Abnormal acuteness of the sense of hearing.
• “The term “hyperacusis” is used to describe the experience of everyday sounds being perceived as intense and overwhelming” (Baguely & Hoare, 2018)
• May cause depression, anxiety, insomnia, suicidal thoughts (Aazh & Moore, 2017).
Thoughts about Suicide and Self-Harm in Patients with Tinnitus and Hyperacusis (Aazh and Moore, 2018)

• “Audiologists offering tinnitus and hyperacusis rehabilitation should screen for suicidal and self-harm ideations among patients, especially for those with comorbid depression, and make onward referral to appropriate services when needed.”

• A total of 150/402 of patients answered the question about suicidal and self-harm ideations (13% positive ideation).
Cause of hyperacusis

- Noise exposure-continuous
- Noise exposure-impulsive
- Hearing loss (sudden)
- Anxiety
- Medications
- Meniere's Disease
- Ear infection
- Aging
- Head or Neck trauma
- Infection/virus
- Depression
- Migraine
- Other

Number of patients

Tyler, 2015
Other Underlying factors include:

- Bell’s Palsy
- Temporomandibular joint problems
- Central Nervous System damage
- Meniere’s Disease
- Hypothyroidism
- Lyme Disease
- PTSD
- Depression
- Migraine Headaches
- Superior Canal Dehiscence (SCD)
- Lateral Canal Dysplasia (LCD)
- ...

...
Disorders comorbid with hyperacusis

- Posttraumatic stress disorder
- Chronic fatigue syndrome
- Generalized anxiety disorder
- Depression
- Exhaustion
- Fibromyalgia
- Irritable bowel syndrome
- Migraine
- hearing impairment
- Tinnitus
- Back/joint/muscle disorders

Paulin, Andersson, Nordin (2016)
Classification of hyperacusis
(Tyler et al., 2014)

• Loudness Hyperacusis
• Annoyance Hyperacusis
• Fear Hyperacusis
• Pain Hyperacusis
Prevalence of Hyperacusis

• In general public, it is unknown. Estimates of 10-15%.
• 18% in children with autism (N=199) (Rosenhall et.al, 1999)
• 46% in a pediatric otolaryngology clinic (Coelho & Sanchez, 2004)
• Up to 40% of children with autism (Autism Research Institute)
• Smaller auditory dynamic ranges in autistic group associated with increased perception of loudness (Khalfa et al., 2004)
• 95% in children with Williams syndrome (Borse, Curfs, & Fryns, 1997)
• 69% in individuals with Asperger’s Syndrome (Danesh, et al, 2015)
Understanding Neural bases of Hyperacusis (emphasis on cochlear synaptopathy)

Image Source: https://hyperacusisresearch.org/2017-aro-hyperacusis-next-steps/
“Hidden” hearing loss

• Loss of synaptic connections between inner hair cell and the dendritic processes of spiral ganglion cells (Kujawa and Liberman, 2009).

• Can occur without elevation of auditory thresholds (as measured in standard audiometric evaluation). Significantly impacts high threshold, low spontaneous firing rate cochlear neurons.

• Synaptic connections appear to be the most at-risk elements of the peripheral auditory system, and their loss may lead to subsequent hair cell death and/or cause central auditory system hyperactivity (Liberman and Kujawa, 2017).
Hyperacusis and Cochlear Synaptopathy

- Noise exposed mice (neuropathic noise group, nonneuropathic noise group, and control) displayed hypersensitivity to sound via an acoustic startle reflex paradigm.

Hickox and Liberman (2014)
Hyperacusis and Cochlear Synaptopathy

Hickox and Liberman (2014)
Hyperacusis and Cochlear Synaptopathy

Hickox and Liberman (2014)
Hyperacusis and Cochlear Synaptopathy

Hickox and Liberman (2014)
Central gain control in tinnitus and hyperacusis

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Sensorineural hearing loss induced by noise or ototoxic drug exposure reduces the neural activity transmitted from the cochlea to the central auditory system. Despite a reduced cochlear output, neural activity from more central auditory structures is paradoxically enhanced at suprathreshold intensities. This compensatory increase in the central auditory activity in response to the loss of sensory input is referred to as central gain enhancement. Enhanced central gain is hypothesized to be a potential mechanism that gives rise to hyperacusis and tinnitus, two debilitating auditory perceptual disorders that afflict millions of individuals. This review will examine the evidence for gain enhancement in the central auditory system in response to cochlear damage. Further, it will address the potential cellular and molecular mechanisms underlying this enhancement and discuss the contribution of central gain enhancement to tinnitus and hyperacusis. Current evidence suggests that multiple mechanisms with distinct temporal and spectral profiles are likely to contribute to central gain enhancement. Dissecting the contributions of these different mechanisms at different levels of the central auditory system is essential for elucidating the role of central gain enhancement in tinnitus and hyperacusis and, most importantly, the development of novel treatments for these disorders.

Keywords: tinnitus, hyperacusis, central gain enhancement, lateral inhibition, homeostatic plasticity
Identifying the Neural Network(s) for Hyperacusis (Salvi, ARO, 2017)
Pain and hyperacusis nociception

- Type II neurons innervating OHCs in the cochlea mediate auditory nociception.

Underlying Pathologies
Hyperacusis and Ménière’s Syndrome

• Ménière’s syndrome involves severe dizziness, tinnitus and hearing loss. These symptoms primarily result from increased endolymph pressure in the cochlear and vestibular canals.

• Some studies have reported patients with hyperacusis and Ménière’s. In a clinic study of 102 patients with Ménière’s, it was reported that a louder tinnitus had a correlation with more severe hyperacusis and hearing loss, but not with vertigo.

Tyler et al. (2014)
Hyperacusis Following Unilateral Damage to the Insular Cortex: A Three-Case Report

• Boucher et al (2015) reported three cases who developed hyperacusis following insular cortex damage.

• Two patients had had an isolated unilateral insular stroke and a patient with epilepsy who received a right insular resection in order to control seizures.

• All patients, displayed loudness discomfort levels that were lower than typical.

Boucher et al. (2015)
Hyperacusis / Insular Lobe Lesion

MRI scans of each patient

Boucher et al. (2015)
A group of researchers described a case of middle cerebral aneurysm that presented with brief, intermittent episodes of bilateral hyperacusis. Audiologic and otologic examinations were conducted, however the results were normal and no tinnitus was present.

Khalil et al. 2002, Cited in Tyler et al. (2014)
Hyperacusis and Autism

• “Autism may include the symptoms of exaggerated reactions to vision, hearing, touch, smell, or taste. This condition can affect social and communicative development and several studies have linked it to hyperacusis.” Tyler et al. (2014)

• Sound hypersensitivity in the autism group may be due to abnormality of the efferent auditory pathway as shown by lack of sufficient contralateral suppression. Danesh & Kaf (2012)
Hyperacusis & Bell's palsy (30%)

Cranial Nerves that may be associated with decreased sound tolerance

1. Olfactory
2. Optic
3. Occulomotor
4. Trochlear
5. Trigeminal
6. Abducens
7. Facial
8. Vestibulo<br>(Vestibulocochlear)
9. Glossopharyngeal<br>Vagus
10. Spinal accessory/Accesory
11. Hypoglossal

On Old Olympus' Towering Top, A Finn And German Viewed Some Hop
Tonic tensor tympani syndrome

When the tensor tympani contracts without the presence of a loud sound it is referred to as Tonic Tensor Tympani Syndrome (TTTS).
LYME DISEASE

- Flu-Like Symptoms
  - Headache
  - Fatigue
  - Fever
  - Chills
  - Sore Throat
  - Muscle Aches

- Psychological Complications (Long Term)
  - Depression
  - Dementia

- Rash at the Site of the Tick Bite - Itching

ERYTHEMA MIGRANS
Superior Canal Dehiscence and Hyperacusis

Superior semicircular dehiscence syndrome (SCD) refers to the partial absence of bone covering the superior canal in the inner ear, which can affect both hearing and vestibular functions (Minor, 2005).
Superior canal dehiscence
Vestibule and horizontal canal

(This patient is a case who faints after hearing loud sounds such as siren which has developed to decreased sound tolerance issues and avoidance).
Hyperacusis due to lateral semicircular canal involvement
Can Lateral Semicircular Canal Dysplasia Play a Role in the Genesis of Hyperacusis?

• Modugno & Brandolini (2014), studied three cases that were characterized by disabling hyperacusis in which semicircular canal dehiscence (SSCD) was excluded by temporal bone high-resolution computed tomography (HRCT).
• Their study presented three cases with bilateral lateral canal dysplasia (R/O history of head trauma)
Evaluation of Hyperacusis
Loudness Discomfort Levels (LDLs)
(not looking to the level that how much you can tolerate!)

- Typically LDLs can be assessed using stimuli such as pure tones, warble tones, noise and even even speech.
Questionnaires for Hyperacusis

1. Hyperacusis Questionnaire (Khalfa et.al., 2002)
2. Modified Khalfa Hyperacusis Questionnaire

Supplementary Questionnaires:

1. Generalised Anxiety Disorder (GAD-7)
2. Short Health Anxiety Inventory (SHAI)
3. Mini-Social Phobia Inventory (Mini-SPIN)
4. Obsessive Compulsive Inventory-Revised (OCI-R)
5. Panic Disorder Severity Scale-Self Report (PDSS-SR)
6. Patient Health Questionnaire (PHQ-9)
7. Penn State Worry Questionnaire-Abbreviated version (PSWQ-A)
8. Hospital Anxiety and Depression Scale (HADS)
Treatments and management methods

We are hair cells and you have approximately 40,000 of us in your inner ear.

Loud noise gives us a hard time.

Too much noise can kill us, which means hearing loss for you.

Inner ear: hair cells healthy

Hair cells are gone
Psychological Treatments

- Counseling and Education
- Mindfulness Therapy
- Cognitive Behavioral Therapy
- Dialectical Behavior Therapy
Role of patient education
Hyperacusis Treatments: Cognitive Behavioral Therapy (CBT)

• The cognitive behavioral therapy for hyperacusis evolved from the cognitive behavioral therapy for tinnitus and anxiety disorders. Treatment includes education, applied relaxation, graded exposure to sounds, and cognitive therapy for distressing thoughts and beliefs regarding sounds.

Pienkowski et al. (2014)
Hyperacusis Treatments: Cognitive Behavioral Therapy

• RCT with 60 subjects, each reporting hyperacusis as her or his primary audiologic problem.
• Primary outcome measure: loudness discomfort levels (LDL)
• Secondary outcome measures: Hyperacusis Questionnaire (HQ), Hospital Anxiety and Depression Scale (HADS), Tampa Scale for Kinisiophobia (TSK), and The Quality of Life Inventory (QOLI).
• Subjects were randomly assigned to either a treatment group (CBT for hyperacusis) or a waiting-list control group.
Hyperacusis Treatments: Cognitive Behavioral Therapy

- CBT treatment included six therapy sessions consisting of hyperacusis education, individualized questioning about hyperacusis reaction, exposure therapy, environmental sound enrichment, applied relaxation, behavioral activation, and goal-setting (Jüris, Andersson, Larsen, & Ekselius, 2014).
Hyperacusis Treatments: Cognitive Behavioral Therapy

- Significant between group (treatment vs. wait-list control) were detected on each outcome measure (except for the Hospital Anxiety and Depression Scale) in favor of the CBT group.
- Treatment effects were intact at a 12 month follow up evaluation.
- Between group differences were no longer measured when the wait-list control group was treated with the same CBT methodology (Jüris, Andersson, Larsen, & Ekselius, 2014).
Hyperacusis

Treatments: Cognitive Behavioral Therapy

• CBT can be an effective treatment for patients with hyperacusis.
• Noise-related avoidance behaviors need to be targeted in treatment (Jüris, Andersson, Larsen, & Ekselius, 2014).

Fig. 2. Results for the LDL test at the four assessments: 1 = baseline; 2 = post treatment for the CBT group and pre-treatment for the WL group; 3 = post-treatment for the WL group; and 4 = 12-month follow-up for both groups. Significant differences exist between CBT- and WL-groups at assessment 2 for both ears.
Sound Therapy

- Use of music
- Desensitization with pleasant sounds
- Tinnitus Retraining Therapy (modified) (Jasterboff & Jasterboff, 2000)
- Fractal tone Therapy
Hyperacusis Treatments: Sound Therapy

• Studies have found that listening to low-level sounds for several months can alleviate tinnitus and hyperacusis.

• Over time there are gradual increases of the level and/or duration of the sound treatment that are implemented with positive reinforcement from the clinician. The association of positive experiences with sound treatment can result in the elimination of hyperacusis symptoms.

Pienkowski et al. (2014)
Hyperacusis Treatments: Sound Therapy

• In the case of fear hyperacusis, behavioral modification is necessary to reduce aversion to phobic stimuli before loudness desensitization can occur.

• The following are four general sound therapy strategies for hyperacusis:
  – Continuous Low-Level Broadband Noise
  – Successive Approximations to High-Level Broadband Noise
  – Successive Approximations to Troublesome Sounds
  – Gradual Increase of Maximum Output of Hearing Aid

Pienkowski et al. (2014)
Hyperacusis Treatments: Sound Therapy

• “Adaptive plasticity of loudness induced by chronic attenuation and enhancement of the acoustic background” (Formby, Sherlock, & Gold, 2003).
• Researchers indirectly examined central gain hypothesis via controlled alteration of normally hearing subjects acoustic background (using earplugs or low-level sound therapy devices).
• Subjects wore either earplugs or sound generators continuously for two weeks (at least 23 hours a day).
• Pre and post loudness judgments were measured using the Contour Test of Loudness Perception.
Hyperacusis Treatments: Sound Therapy
Hyperacusis Treatments: Sound Therapy

• Treatment effects indicating that a subjects’ chronic acoustic background can modulate her or his loudness perceptions.
• Indirect evidence for compensatory processes of the central auditory pathways (and the plasticity of these pathways).
• In terms of hyperacusis, these results (along with other investigations in the research literature; Noreña & Chery-Croze, 2007; Formby et al., 2015) advocate acoustic environment enrichment and sound therapy to increase loudness tolerance, rather than sound avoidance (via earplug use) which may in fact exacerbate a patient’s problems (Formby, Sherlock, & Gold, 2003).
Medical and Surgical Treatments
Hyperacusis Treatments: Medications

• There is a high interest in using medications to treat hyperacusis, however it has not been investigated in clinical trials. The work published in regard to this is limited to clinical case reports.

Pienkowski et al. (2014)
Hyperacusis Treatments: Medications

• A case study in 1993 described the use of Alprazolam (a short-acting anxiolytic) in 5 patients who presented with tinnitus and hyperacusis. After 8 weeks of treatment a complete remission of hyperacusis was observed.

• Another case study focused on the use of Carbamazepine (an anticonvulsant and mood-stabilizing drug) for the relief of hyperacusis in two patients diagnosed with Lyme disease. A third study described the use of selective serotonin receptor inhibitors (fuvloxamine and fluoxetine) for a patient with complete remission of hyperacusis and increase of ULLs.

Pienkowski et al. (2014)
Round and oval window reinforcement for the treatment of hyperacusis

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Round and oval window reinforcement for the treatment of hyperacusis

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

• Herbert Silverstein, MD, Yi-Hsuan Emmy Wu, MD, & Suzannah Hagan, AuD (2014)
Round and oval window reinforcement for the treatment of hyperacusis

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).

•Herbert Silverstein, MD, Yi-Hsuan Emmy Wu, MD, & Suzannah Hagan, AuD (2014)
Audiometric and loudness discomfort level (LDL) results for Patient 2, right ear.

• Abstract: Hyperacusis is intolerance of certain everyday sounds that causes significant distress and impairment in social, occupational, recreational, and other day-to-day activities. The aim of this report is to summarize the key findings and conclusions from the Third International Conference on Hyperacusis and highlight implications for research and clinical practice.

• The main topics discussed comprise: (1) diagnosis of hyperacusis and audiological evaluations; (2) neuro-biological aspect of hyperacusis; (3) misophonia; (4) hyperacusis in autism spectrum disorder; (5) noise sensitivity; (6) hyperacusis-related distress and co-morbid psychiatric illness; and (7) audiologist-delivered cognitive behavioral therapy for hyperacusis.
Take home message!

• Hyperacusis (a decreased sound tolerance disorder) is a manageable condition!