### Decreased Sound Tolerance Disorders:
What They are and How to Manage Them

<table>
<thead>
<tr>
<th>James W. Hall III, PhD</th>
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<tr>
<td>Professor</td>
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<tr>
<td>Salus University</td>
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<tr>
<td>Adjunct Professor</td>
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<tr>
<td>Nova Southeastern University</td>
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<td>University of Florida</td>
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<tr>
<td>Extraordinary Professor</td>
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<td>University of Pretoria South Africa</td>
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### "Patient" Inquiry via Email (September 14, 2014)

**Dr Hall,**

I am 27. My hyperacusis is at a severe crisis level. I suspect I am a category 4. My tolerances have continued to fall over the last few weeks, just exposed to everyday sound. I would estimate my LDL’s are in the 30’s. It has become very severe in the last 24 hrs. I have limbic/autonomic disfunction with high blood pressure, dizziness upon standing up, lightheadedness, constant on edge feeling, chest pain, anxiety, panic attacks, and I was only sleeping a few hours a night. Last night I did not sleep at all! I have orthostatic hypotension (my blood pressure drops 20 points when I sit up or stand up) and I believe postural orthostatic tachycardia (my heart rate goes up 30 bpm when I sit up or stand up).

Today I have also noticed occasional arrhythmia. My family doctor doesn't have a clue what is going on, she gave me Buspar for the anxiety (which I don't feel like is helping) and Clonidine (lowers blood pressure, not Klonopin the benzo, to help me sleep which is no longer working). I am considering going to the ER but at this point I am afraid that more sound exposure could potentially setting me back even more.

Please advise. I am in Oregon and I have an appt to see TRT specialist Marsha Johnson today, but not sure if I will make it.

Can you advise on medications. Thank you so much for your time.
Dear Mr. _______

Sorry for my delayed response. I'm no longer at the University of Florida, and I don't regularly check my old email account.

I'm sorry to hear about your hyperacusis and its affect on your quality of life. However, I'm confident that Marsha Johnson will help you. I suggest that you contact her as soon as possible to request an earlier appointment. Meanwhile, your intolerance to sound will be helped if you are always surrounded by soft and relaxing background sound.

Feel free to contact me again at my new email address pending your appointment with Dr. Johnson.

Best wishes....
### Terminology and Definitions (1)

- "Consistently exaggerated or inappropriate responses or complaints of uncomfortable loudness to sounds that are neither intrinsically threatening nor uncomfortably loud to a typical person" (Klein et al. Hyperacusis and otitis media in individuals with Williams syndrome. JSHD 55: 1990)
- "Unusual intolerance to ordinary environmental sounds" (Vernon. Pathophysiology of tinnitus: a special case -- hyperacusis and a proposed treatment. Am J Otol 8: 1987)
- "Experience of inordinate loudness of sound that most people tolerate well, associated with a component of distress ... this experience has a physiologic basis ... but it also has a psychological component." (Baguley & Andersson, 2007)

### Terminology and Definitions (2)

<table>
<thead>
<tr>
<th>Hyperacusis</th>
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<tbody>
<tr>
<td>Abnormally strong reactions (intolerance) occurring within the central auditory pathways with exposure to moderate levels of sounds</td>
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<tr>
<td>Hearing sensitivity is typically normal</td>
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<tr>
<td>Otoacoustic emissions (OAEs) are typically normal</td>
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<tr>
<td>Lack of contralateral suppression of OAEs</td>
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<tr>
<td>Reduced LDLs</td>
</tr>
<tr>
<td>Loudness recruitment</td>
</tr>
<tr>
<td>Abnormal growth of loudness</td>
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<tr>
<td>Sensory (outer hair cell) hearing loss (peripheral)</td>
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<tr>
<td>OAEs are abnormal</td>
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<tr>
<td>Reduced LDLs</td>
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### Terminology and Definitions (3)

<table>
<thead>
<tr>
<th>Phonophobia</th>
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<tr>
<td>Fear of sound</td>
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<tr>
<td>Anticipatory, anxious, and sometimes different reaction to specific sounds, e.g.,</td>
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<tr>
<td>Vacuum cleaner</td>
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<td>Telephone ringing</td>
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<tr>
<td>Crying child</td>
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<tr>
<td>Reaction and avoidance to sounds involves learning and conditioning</td>
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<tr>
<td>Consistently related to intensity of sound</td>
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<tr>
<td>Intact peripheral and efferent auditory systems</td>
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<tr>
<td>Enhanced connections between the auditory cortical regions and the limbic and autonomic nervous system regions</td>
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</table>
## Decreased Sound Tolerance Disorders: What They Are and How to Manage Them: Terminology and Definitions (4)

- Misophonia or Selective Sound Sensitivity Syndrome (SSSS or 4S)
  - Questions about whether it is auditory or non-auditory and physiological or psychological
  - Dislike or hatred of sound (may be selected sounds)
  - Irritation and dislike of specific soft sounds
  - Most common sounds are repetitive, perceived as unpleasant, produced by family members, e.g.,
    - Eating
    - Smacking lips
    - Breathing
    - Speech sounds

## Misophonia: Selected Readings (1)


## Misophonia: Selected Readings (2)

Misophonia: 
Selected Readings (3)

- Cavanna AE (2014). What is misophonia and how can we treat it? *Expert Rev Neurother*, 14, 357-359

Review Article on Misophonia
*(Spankovich & Hall, Audiology Today (July/August), 2014)*

Decreased Sound Tolerance Disorders: 
What They are and How to Manage Them: 
*Good Reference*

Decreased Sound Tolerance Disorders:
What They are and How to Manage Them

- Different categories of decreased sound tolerance
- Definitions
- Disorders associated with the symptom of hyperacusis
- Mechanisms of hyperacusis
- Assessment of hyperacusis
- Management options for hyperacusis and other forms of decreased sound tolerance
- Illustrative case reports

Decreased Sound Tolerance Disorders:
Hyperacusis Mechanisms

- Imbalance of neuro-chemical serotonin (5 HT), involved in
  - Stimulus reactivity
  - Perception of sensory information in brain
- Release of neurotransmitter glutamate with stress, anxiety, or fatigue, e.g., inner hair cell synapse with afferent auditory nerve fibers
- Defective efferent (descending or inhibitory) auditory system (specifically medial efferent pathways)
- Increased "central gain" in auditory system
- Activation of limbic system (amygdala) involved in fear conditioning
- Neural spread from auditory system (e.g., lateral lemniscus or thalamus to central trigeminal pathways)... explanation for perception of pain response to sound
- Point prevalence may be as high as 8 to 9% (e.g., Andersson et al. Hypersensitivity to sound (hyperacusis): a prevalence study conducted via the internet and post. Int J Audiology 41: 2002.

Hyperacusis

- Increased central gain
- Likely auditory efferent system involvement
- Interaction between auditory cortex and
  - Limbic system
  - Autonomic nervous system
  - Pain centers
- Multiple etiologies
- Audiologists can offer management options
HYPERACUSIS: Three Factors Influencing Hearing Sensitivity or "Gain" (Adapted from Baguley and Andersson, 2007)

- Amount of sound in the environment is monitored by the nervous system. Sensitivity is adjusted when a person is in the presence of a loud sound.
- The mood or emotional state of a person influences auditory gain. Sounds are perceived as more intense than usual for a person who is anxious or frightened. Such a person might even startle for everyday sounds, e.g., a telephone or doorbell.
- The meaning of sound, how easily it is remembered and interpreted and how loud it is perceived, can influence hearing sensitivity.

Pawel Jastreboff, PhD
"Neurophysiological Model of Tinnitus and Hyperacusis"

Hyperacusis Anatomy & Physiology

Auditory region of the brain (Heschl’s gyrus) in the temporal lobe of auditory cortex ... responsible for decreased sound tolerance
Limbic System

Emotional center of the brain

Autonomic Nervous System

Controlling the brain’s response to “danger sounds”

Hyperacusis and Other Decreased Sound Tolerance Disorders

- Different categories of decreased sound tolerance... definitions
- Disorders associated with the symptom of hyperacusis
- Mechanisms of hyperacusis
- Assessment of hyperacusis
- Management options for hyperacusis and other forms of decreased sound tolerance
- Illustrative case reports
Hyperacusis and Other Decreased Sound Tolerance Disorders: Assessment

- CONSULTATION
- AUDIOLOGIC ASSESSMENT
- REFERRAL FOR EVALUATION OR Rx
- AUDIOLOGIC MANAGEMENT

Diagnostic Hyperacusis Interview
(Adapted from Baguley & Andersson, 2007)

- Background questions
  - Family situation
  - Work situation (current and past)
  - Sick leave?
  - Compensation?
  - Legal action?
- Noise sensitivity questions
  - Onset; gradual or sudden
  - Types of aversive sounds
  - Reactions to sounds
    - Fear
    - Annoyance
    - Other

Diagnostic Hyperacusis Interview (2)
(Adapted from Baguley & Andersson, 2007)

- Medical history
  - Depression ... before or after onset of hyperacusis?
  - Consultation with psychiatry or psychology?
  - Migraine?
  - Use of ear protection?
  - Medications ... list (associated with hyperacusis?)
  - Other sensitivities or medical problems?
    - Light
    - Touch
    - Pain
    - Smell
    - Allergy
    - Balance
    - TMJ disorders
Hyperacusis and Other Decreased Sound Tolerance Disorders: History

- Medical history, e.g.,
  - Neurological disorders or insult
  - Chronic otitis media
  - Psychological disorders
  - William’s syndrome
  - Head injury
  - Migraine headaches
  - Multiple sclerosis?
- Family history of sensory hypersensitivity?

Hyperacusis and Other Decreased Sound Tolerance Disorders: History

- Audiologic history, e.g.,
  - Infant risk indicators
  - Previous audiograms
  - CAPD
  - Tinnitus
- Other related disorders, e.g.,
  - Sensory integration disorder
  - Autism spectrum disorders?

Hyperacusis and Other Decreased Sound Tolerance Disorders: History

- Description of complaints
  - Sounds that are bothersome or intolerable
  - Sounds that are pleasant or tolerable
  - Hyperacusis vs. phonophobia?
  - Maturational vs. disordered sound intolerance
- Reaction to sounds that are bothersome or intolerable
  - Covering ears
  - Use of earplugs and/or earmuffs?
  - Avoidance of noisy places (alteration of daily activities)
  - Running away or potentially dangerous evasive actions
Hyperacusis and Other
Decreased Sound Tolerance Disorders:

**History**
- Attempt to answer all parent/child questions
- Definition and validation of hyperacusis (it does exist!)
- Written information on hyperacusis
- Proceed with further assessment and/or management now or later?
  - Quantify and qualify impact on quality of life
  - Avoidance of social and important activities, e.g.,
    - School
    - Work
    - Past-times (e.g., music, sports events, etc)
- Full written report for parents and others as requested

**Summary of History**
- A complete history
  - Is essential for understanding a patient's history and developing an appropriate management plan
  - Is important in making referral decisions
  - Permits interactions with a patient that facilitate the trusting professional relationship needed for effective management
- Inventories quantify impact on quality of life
- A thorough diagnostic assessment provides clues about the origin and nature of tinnitus and/or hyperacusis
- Counseling opportunities during the history and assessment contribute importantly to minimizing impact of tinnitus/hyperacusis on quality of life

**Assessment**
- Protocol for Auditory Assessment (< 30 minutes)
  - Inform patient that any test can be terminated if any of the sounds are too loud
  - Impittance measurement
    - Tympanometry
    - No acoustic reflexes!
  - Pure tone audiometry
    - Thresholds measured to as low as – 10 dB HL
    - Inter-octaves > 2000 Hz
    - High frequency thresholds up to 20,000 Hz
    - Bone conduction only as indicated
Protocol for Auditory Assessment (< 30 minutes)

- Distortion product otoacoustic emissions (DPOAE)
  - Frequencies from 500 to 8000 or 10,000 Hz
  - 6 or 8 frequencies per octave
  - 65/55 intensity levels
  - Ask patient to raise hand if they hear the tinnitus pitch

- Speech audiometry
  - Measure at a comfortable level
  - NU-6 lists with most difficult words ranked from 1 - 25

- Loudness discomfort levels (LDLs)
  - Pure tones and speech

DPOAEs are Invaluable in the Assessment and Management of Hyperacusis and Disorders of Sound Tolerance

Tinnitus Handicap Inventory
Can be Adapted for Hyperacusis

- 25 items (see handout)
  - 12 on functional subscale, e.g.
    - "Because of your tinnitus do you have trouble falling to sleep at night?"
  - 8 on emotional subscale, e.g.
    - "Does your tinnitus make you angry?"
  - 5 on catastrophic subscale
    - "Do you feel that you cannot escape your tinnitus?"

Three subscales
- 12 questions in functional (F) subscale, e.g.,
  - “Because of your tinnitus do you have trouble falling to sleep at night?”
- 8 questions on emotional (E) subscale, e.g.,
  - “Does your tinnitus make you angry?”
- 5 questions on catastrophic (C) subscale, e.g.,
  - “Do you feel that you cannot escape your tinnitus?”


Three options for answering questions
- Yes = 4 points
- Maybe = 2 points
- No = 0 points
- 100 point total (most severe impact on quality of life)


Grade Score
Grade 1: No Handicap 0 - 16
Grade 2: Mild Handicap 18 - 36
Grade 3: Moderate Handicap 38 - 56
Grade 4: Severe Handicap 58 - 76
Grade 5: Catastrophic Handicap 78 - 100

Handscomb (2006)
McCombe, et al. (2001)
Decreased Sound Tolerance Disorders:
What They are and How to Manage Them

- Different categories of decreased sound tolerance definitions
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- Illustrative case reports

Patient/Family Counseling and Education in Disorders of Sound Tolerance:
Counseling is Intervention

“Knowledge is power.”
(Nam et ipsa scientia potestas est.)

Francis Bacon
(1561-1626)

Meditationes Sacrae (1597)

Recent Publication (JWH3)

AudiologyOnline
200: Treating Patients with Hyperacusis and Other Forms of Decreased Sound Tolerance
James W. Hall, MD, PhD
20091030

From the desk of Gus Mueller
Decreased Sound Tolerance Disorders: What They are and How to Manage Them
(The Hearing Journal, 51, April, pp 10-15, 1998)

General Counseling Guidelines (1)

- Conduct counseling in a quiet and private venue
- Introduce yourself with eye contact and a firm (but comfortable) handshake
- Use respectful titles (e.g., Mr. or Mrs.)
- Give patient and family members the impression that you have plenty of time and nothing is more important than talking with them
- Be accepting of family statements, feelings, and attitudes but...
- Gently clarify any misconceptions with factual information

General Counseling Guidelines (2)

- Explain test findings slowly and simply, without jargon
- Emphasize positive or encouraging findings
- Pause frequently during explanation to allow for spontaneous questions
- Be sensitive, understanding, and compassionate
- Before meeting ends, ask patient and family members if they have any questions
- Ask patient for permission to release the formal written report to the primary care physician and to the patient (if permission is required)
### Decreased Sound Tolerance Disorders: What They are and How to Manage Them

#### Be a Good Listener (1)
- Be an active listener
- Ask open ended questions, e.g. “Tell me about your concerns regard the tinnitus you hear.”
- Ask family members to clarify or repeat what they said to find out “where they are coming from”
- Listening doesn’t imply agreement with what a person is saying
- Good listening is not easy or natural
- Don’t plan your response or your solution to the problem while a person is still talking (I’m not necessarily preaching what I always practice!)

#### Decreased Sound Tolerance Disorders: What They are and How to Manage Them

#### Be a Good Listener (2)
- Really pay attention to the patient or family member and to what he/she is saying… don’t just pretend
- Focus on content, not style, when listening to family members
- Don’t “multi-task” while listening (e.g., completing forms, taking notes, looking at cell phone)
- Don’t react negatively to what the family member says

#### Counseling in Decreased Sound Tolerance Disorders: Addressing the Limbic System
- Normal patient family emotional reactions or responses to bothersome tinnitus
  - Shock, Confusion, Distress
  - Hopelessness, Frustration
  - Fear, Anxiety, Worry
  - Anger, Bitterness, Insecurity
  - Denial, Guilt, Depression
Amygdala responses interfere with frontal lobe responses, including:
- Reasoning
- Learning
- Remembering
- Understanding

Decreased Sound Tolerance Disorders: 
Addressing the Limbic System (2)

Counseling Patients and Family Members:
The Placebo Effect is Your Friend

Hyperacusis (and Tinnitus) Research
- Placebo effect is a very clear factor in outcome
- Accounts for up to 40% of apparent benefit from “treatment”
- Control (non-treatment) group is essential in hyperacusis and tinnitus research

Clinical Service
- Placebo effect can contribute significantly to improved patient outcome
- Placebo effect is enhanced by:
  - Perceived and actual expertise of the audiologist
  - Good bedside manner
  - Confident and compassionate interactions with patient

Sound Enrichment in the Management of Decreased Sound Tolerance Disorders

<table>
<thead>
<tr>
<th>Environment Sound Machine</th>
<th>Sound Pillow</th>
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<tr>
<td>~ $25</td>
<td>~ $30</td>
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Sound Enrichment is a Simple and Generic Version of Tinnitus Retraining Therapy (TRT)

- **Directive Counseling**
  - Educational counseling that helps patient *neutralize* the negative *reaction* to tinnitus
  - Describes tinnitus mechanisms to *rationalize* and *demystify* the tinnitus

- **Sound Therapy**
  - Helps patient *minimize* tinnitus *perception*
  - Uses soft background noise without completely masking the tinnitus
  
  Jastreboff (1990)

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Pawel Jastreboff, Ph.D.
Originator of “Tinnitus Retraining Therapy (TRT)”

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Tinnitus Retraining Therapy: The “Neurophysiologic Model” is Also Effective for Hyperactive

*The model stresses (a) the importance of the basic principles of the functioning of the nervous system, such as the capacity to be habituated to signals that are emotionally neutral and do not carry important information, and (b) the fundamental feature of the nervous system -- its plasticity.*

Jastreboff, Gray & Gold, 1996
Tinnitus Retraining Therapy (TRT)

- Directive Counseling
  - Educational counseling that helps patient *neutralize* the negative *reaction* to tinnitus
  - Describes tinnitus mechanisms to *rationalize* and *demystify* the tinnitus

- Sound Therapy
  - Helps patient *minimize* tinnitus *perception*
  - Uses soft background noise without completely *masking* the tinnitus

Jastreboff (1990)

Neurophysiological Model for Tinnitus Retraining Therapy (TRT)

Hundreds of Drugs May Enhance or (Rarely) Produce Hyperacusis (and Tinnitus) for A Small Minority of Patients Based on Clinical Trials
Neuromonics Treatment for Hyperacusis:
www.neuromonics.com

Haven™ Device: “Blends an individualized, high-frequency signal with relaxing music to provide patients an on-demand sense of control and relief while wearing the device.”

Oasis™ Device: “An FDA-cleared, patented and clinically proven device that offers long-term treatment and significant relief for severe tinnitus.”

Sanctuary ™ Device: “Functions in a similar manner as the Haven, providing situational relief, but works with a choice of pre-programmed profiles rather than individualized spectral profiles.”

Neuromonics Tinnitus Treatment:
www.neuromonics.com

Step 1: Comprehensive Personalized Assessment
An audiologist conducts a thorough hearing (up to 12.5kHz) and tinnitus assessment, discusses tinnitus history and provides some education about tinnitus and its causes. Once the hearing and tinnitus profile is determined, recommendations are made about the most appropriate treatment options including the Neuromonics Tinnitus Treatment.”
Neuromonics Tinnitus Treatment:
Five Step Treatment

“Step 2: Processor Configuration
Each individual’s audiometric profile is used to make their prescription for their Neuromonics Processor. The Neuromonics device is small and lightweight (it's about the same size and weight of a cell phone) and accompanied by high fidelity earphones. It delivers the acoustic component of the treatment.

The acoustic stimulus provides stimulation across a considerably wider range of frequencies than other available treatments, up to 12.5 kHz. The acoustic signals are distinct for each ear and correlated, and are provided in stereo. This further ensures stimulation of the multiple ipsilateral and contralateral pathways of the auditory system.

Neuromonics Tinnitus Treatment:
Five Step Treatment

“Step 3: Pre-Conditioning Stage
During the Pre-Conditioning Stage, users are advised to listen to their device for at least two hours each day. In this initial stage, the device delivers an acoustic neural stimulus that provides a high level of interaction with their tinnitus perception. The benefits of this stage are relaxation, sleep and relief from tinnitus symptoms. Support and education is provided throughout this stage by an Audiologist.

Neuromonics Tinnitus Treatment:
Five Step Treatment

“Step 4: Active Treatment Stage
During this Stage, users also listen to the device for at least two hours per day. At this stage, an acoustic neural stimulus provides a lower level of interaction. Intermittent interaction with the tinnitus perception facilitates the desensitisation process, i.e., reprograms the brain to filter out the sound of the tinnitus. This process has proven to be efficient and effective at reducing the associated disturbance and impact on quality of life.”
### Neuromonics Tinnitus Treatment:
**Five Step Treatment**

**“Step 5: Maintenance Stage**
After a successful program of treatment lasting around six months, a maintenance program is devised for each individual to manage and control their tinnitus independently of clinician support. Many users find they may not feel the need to use the device any more. If they do continue listening, it’s usually for less often and for less time each day to maintain the benefits achieved.”

### Decreased Sound Tolerance Disorders:
**Hyperacusis is a Symptom Associated with Varied Clinical Entities (1)**

- Central neurological disorders
  - Depression
  - Migraine
  - Chronic fatigue syndrome
  - Post-traumatic stress disorder
  - Tay Sach’s disease
  - Ramsay-Hunt syndrome
  - Multiple sclerosis
  - Middle cerebral artery aneurysm
  - Complex regional pain syndrome related dystonia

- Lyme disease
- Facial paralysis
- Pyridoxine deficiency
- Benzodiazepine dependency
- William’s syndrome
- Autism
## Williams Syndrome

(www.williams-syndrome.org)

- Identified in the early 1960s
- Incidence of 1 in 20,000 live births
- Caused by micro-deletion on chromosome 7q11.23, including ~20 genes
- From 50 to 90% of children with WS have hyperacusis
- Features include:
  - Facial features
  - Cognitive deficits, e.g.,
    - Conceptual reasoning
    - Problem solving
    - Arithmetic ability
    - Spatial cognition
  - Fears and anxieties
  - Motor control problems
  - Cardiac abnormalities
  - Language impairment
  - Middle ear disease (otitis media)

## Decreased Sound Tolerance Disorders:

### Hyperacusis is a Symptom Associated with Varied Clinical Entities (3)

- Tinnitus
- Acoustic trauma
- Auto-immune disorders
- Post otologic surgery
  - Ventilation tubes
  - Otosclerosis
  - Tympanoplasty
- Genetic predisposition (family trait)
- Auditory processing disorders (APD)
- Drugs, e.g.,
  - Effexor
  - Prozac
  - Remeron
  - Tegretol
  - Zoloft

## Decreased Sound Tolerance Disorders:

### A Management Team

- Audiologist
  - Assessment and management of hyperacusis is within scope of practice
  - Up-to-date knowledge of hyperacusis is important
  - Credentials should include post-graduate hyperacusis CE activities
  - Knowledge and skills in counseling
  - Professional confidence and compassion
- Primary care physicians, especially pediatricians
  - Audiologist must educate PCPs about hyperacusis
  - PCPs must refer to audiologists all patients with decreased tolerance to sound
Decreased Sound Tolerance Disorders: A Management Team (2)

- Otolaryngologist
  - Preferably an otologist or neurotologist
- Occupational therapist
  - Knowledge and skills in decreased sound tolerance and sensory integration disorder
- Neurologist
  - Knowledge of hyperacusis as symptom of CNS diseases
- Psychologist
  - Offers cognitive behavioral therapy (CPT)
- Psychiatrist

Decreased Sound Tolerance Disorders: Hyperacusis Prevalence in General Population

- Rubinstein et al (1996): 22% (no definition given)
  - Prevalence of 8% (postal survey)
  - Prevalence of 9% (internet survey)
  - Severe hyperacusis estimated in 2 to 3%

Decreased Sound Tolerance Disorders: Hyperacusis Prevalence in Pediatric Population

- Coelho, Sanchez & Tyler (2007)
  - Prevalence in 506 school age children
  - Hyperacusis defined by
    - Questionnaire
    - LDLs
  - Findings
    - 42% of group were bothered by sounds
    - 3.2% met definition of hyperacusis (lowest 5%ile for LDLs)
    - Phonophobia experienced by 9% of children
  - Prevalence of 8% (postal survey)
  - Prevalence of 9% (internet survey)
  - Severe hyperacusis estimated in 2 to 3%
HYPERACUSIS: Sounds that are most bothersome (various sources plus University of Florida clinic)

- Shrii l sounds
- Power saw
- Telephone ringing
- Vacuum cleaner
- Hair dryer
- Sirens (e.g., ambulance)
- Children crying and screaming

Patient/Family Counseling and Education

“Knowledge is power.”
(Nam et ipsa scientia potestas est.)

Francis Bacon
(1561-1626)

*Meditationes Sacrae [1597]*

Primary Management Options for Hyperacusis

- Desensitization
  - Identify bothersome or fearful sounds
  - Suggestions for home management, e.g., honest discussions about bothersome sounds
  - Tape record sounds or download examples from internet
  - Replay the sounds 10 minutes/day louder each day
  - Avoid silence
    - Environmental sound therapy
    - No earplugs unless indicated by behavior and/or noise levels
    - Then use musician earplugs or Doc’s Pro Plugs
Primary Management Options for Hyperacusis

- Explain to patient or parent extended management options
  - Retraining therapy (TRT)
    - Directive counseling
    - Noise generator fitting
  - Neuromonics Tinnitus Treatment
- Referral to other professionals as indicated
  - Occupational therapist (OT)
  - Neurologist
  - Otolaryngologist
  - Psychologist or psychiatrist

HYPERACUSIS: Evidence in Support of Management with TRT

- Bartnik, Fabijanska & Rogowski (1999)
  - Over two-thirds of hypercusis patients showed improvement with TRT
- Gold, Frederick & Formby (1999)
  - Increased LDLs and dynamic ranges for 123 adults with hyperacusis
  - Sound therapy did not eliminate patient concerns about hyperacusis
- Wolk & Seefeld (1999)
  - Positive outcomes with TRT for 23 subjects with troublesome hyperacusis
  - Defined by LDLs, dynamic ranges, and subjective descriptions

HYPERACUSIS: Evidence in Support of Management with TRT

- Jastreboff and Hazell (2004)
  - Summarized existing published and unpublished research
  - Patients with hyperacusis and tinnitus showed greater benefit from TRT than patients with tinnitus only
  - "A significant improvement in hyperacusis patients with TRT has already been reported", however ...
  - No strong clinical evidence for this conclusion was cited from peer-reviewed publications (mostly tinnitus conferences)
- Randomized, double-blind, placebo-controlled clinical trial of efficacy of TRT
- Treatment included counseling and sound therapy with noise generators (NG)
- Treatment administered for > 5 months
- Outcome measured by
  - LDLs
  - Contour Test for Loudness
- Subjects assigned randomly to four treatment groups
  - Full treatment, both counseling and NGs
  - Counseling and placebo NGs
  - NGs without counseling
  - Placebo NGs without counseling.
- Outcome measured by
  - Increase in > 10 dB in LDLs and Contour Test for Loudness
- Subjects assigned to partial treatment group did not benefit from treatment

Madeira, Montmirail, Decat, Gersdorff (2007) Belgium
- TRT investigation
- 24 patients with hyperacusis (out of 46 with tinnitus)
- Sound therapy for minimally 8 hours per day
- Outcome (based on "subjective testimony")
  - Hyperacusis with or without tinnitus, i.e., PJ category 3 (N = 16): 88.5% improved
  - Hyperacusis with or without tinnitus exacerbated by noise, i.e., PJ category 4 (N = 8): 75% improved

- Treatment was enriched sound environment
- Sound therapy administered for less than one month
- Stimuli initially considered "too loud" were perceived as comfortable with > 2 weeks of sound enrichment

Primary Management Options for Hyperacusis:

Conclusions

- Hyperacusis is real
- Hyperacusis can have a major impact on quality of life
- Audiologists are the professionals who should evaluate and manage patients with hyperacusis
- Assessment of hyperacusis should include DPOAEs and high frequency audiometry
- Management should include
  - Proper referrals to other specialists
  - In depth counseling with accurate information
  - Environmental sound enrichment
  - A desensitization program
  - Extended treatment in selected cases
Different categories of decreased sound tolerance ...
definitions
Disorders associated with the symptom of hyperacusis
Mechanisms of hyperacusis
Assessment of hyperacusis
Management options for hyperacusis and other forms of
decreased sound tolerance
Illustrative case report

CASE REPORT:
Hyperacusis in Young Adult

18 year old female
Freshman at the University of Florida (music major)
Referred by out of town otolaryngologist
History
- Onset 10 months earlier when reportedly "damaged ears in loud
  recording studio"
- Hyperacusis is worse in morning
- Cannot tolerate everyday environmental sounds
  - Roommate and friends who laugh and talk loudly
  - Public settings (e.g., restaurants, classroom)
- "General anxiety" for many years
- Xanax in morning and night since onset of hyperacusis for extreme
  anxiety
- Patient repeatedly told she has sensorineural hearing loss

CASE REPORT: Hyperacusis in Young Adult
Initial Consultation
Observations
- Patient accompanied by mother
- Patient clearly anxious
- Crying as relating history and concerns about changing school and
career plans
Questionnaire
- Aware of hyperacusis 100% of waking hours
- On a scale of 0 to 10, effect of hyperacusis on life = 8
- Hyperacusis affects
  - Concentration
  - Sleeping
  - Social events
  - Concerts
Tinnitus Handicap Inventory (THI) at baseline (before consultation)
- Total score = 88
CASE REPORT: Therapy for Hyperacusis in Young Adult

Audiogram

<table>
<thead>
<tr>
<th></th>
<th>Right Ear</th>
<th>Left Ear</th>
</tr>
</thead>
<tbody>
<tr>
<td>0.5</td>
<td>10</td>
<td>10</td>
</tr>
<tr>
<td>1</td>
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<td>10</td>
</tr>
<tr>
<td>2</td>
<td>10</td>
<td>10</td>
</tr>
<tr>
<td>3</td>
<td>10</td>
<td>10</td>
</tr>
<tr>
<td>4</td>
<td>10</td>
<td>10</td>
</tr>
<tr>
<td>6</td>
<td>10</td>
<td>10</td>
</tr>
<tr>
<td>8</td>
<td>10</td>
<td>10</td>
</tr>
</tbody>
</table>

CASE REPORT: Hyperacusis in Young Adult

High Frequency Audiometry

<table>
<thead>
<tr>
<th>Frequency (Hz)</th>
<th>Right Ear</th>
<th>Left Ear</th>
</tr>
</thead>
<tbody>
<tr>
<td>10,000</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>12,500</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>14,000</td>
<td>0</td>
<td>5</td>
</tr>
<tr>
<td>16,000</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>18,000</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>20,000</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

CASE REPORT: Hyperacusis in Young Adult

(L1 = 65 dB SPL; L2 = 55 dB SPL; f2/f1 = 1.2; 5 freq/octave)

<table>
<thead>
<tr>
<th>Frequency (Hz)</th>
<th>DP Amplitude in dB SPL</th>
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<tbody>
<tr>
<td>.50</td>
<td>10</td>
</tr>
<tr>
<td>1K</td>
<td>20</td>
</tr>
<tr>
<td>2K</td>
<td>30</td>
</tr>
<tr>
<td>3K</td>
<td>10</td>
</tr>
<tr>
<td>4K</td>
<td>0</td>
</tr>
<tr>
<td>6K</td>
<td>-10</td>
</tr>
<tr>
<td>8K</td>
<td>-20</td>
</tr>
</tbody>
</table>
CASE REPORT: Hyperacusis in Young Adult

Loudness Discomfort Levels (LDLs)

<table>
<thead>
<tr>
<th>Frequency (Hz)</th>
<th>Right Ear</th>
<th>Left Ear</th>
</tr>
</thead>
<tbody>
<tr>
<td>1000</td>
<td>70</td>
<td>70</td>
</tr>
<tr>
<td>2000</td>
<td>75</td>
<td>75</td>
</tr>
<tr>
<td>4000</td>
<td>80</td>
<td>80</td>
</tr>
<tr>
<td>6000</td>
<td>75</td>
<td>75</td>
</tr>
<tr>
<td>8000</td>
<td>70</td>
<td>80</td>
</tr>
<tr>
<td>Speech</td>
<td>75</td>
<td>65</td>
</tr>
</tbody>
</table>

Initial Management

- Counseling
  - Explanation of test findings
  - Normal hearing (repeated reassurance)
  - Normal cochlear function
  - Documented intolerance to loud sounds
  - Written information about hyperacusis
  - Musician earplugs when exposed to high intensity sound

- Sound enrichment
  - Purchase and use regularly environmental sound device
  - Use iPod at low comfortable level as often as desired
  - Resume normal schedule without worries about hearing

- Return in 4 weeks for follow up visit

Follow Up Consultations at 4 Weeks and 1 Year

- Patient came to clinic unaccompanied
- Patient reported
  - Following all recommendations (has used sound device daily)
  - Tolerance of everyday sounds (including room mate and friends)
  - Happy for first time (since year before initial visit)
  - No longer anxious
- Tinnitus Handicap Inventory
  - One month follow up visit = 10
  - One year follow up visit = 12
- Return only if residual concerns
**CASE REPORT:**
*Hyperacusis in Young Adult*

<table>
<thead>
<tr>
<th>Frequency (Hz)</th>
<th>Right Ear</th>
<th>Left Ear</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Initial</td>
<td>1 Year</td>
</tr>
<tr>
<td>1000</td>
<td>70</td>
<td>95</td>
</tr>
<tr>
<td>2000</td>
<td>75</td>
<td>95</td>
</tr>
<tr>
<td>4000</td>
<td>80</td>
<td>100</td>
</tr>
<tr>
<td>8000</td>
<td>70</td>
<td>&gt; 90</td>
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</table>

- Hyperacusis is real and can have a major impact on quality of life
- Audiologists are the professionals who should evaluate and manage patients with hyperacusis
- Hyperacusis may be a symptom or characteristic of a number of serious neurological and other medical diseases and disorders
- Assessment of hyperacusis should include DPOAEs and high frequency audiometry
- Management should include
  - Proper referrals to other specialists
  - In depth counselling with accurate information
  - Environmental sound enrichment
  - A desensitization program
  - Extended treatment in selected cases not responding to initial management program
  - AIT and other "listening therapies" cannot be recommended due to lack of evidence based research

**Update on the Assessment and Management of Hyperacusis:**
*A Serious and Not Uncommon Auditory Disorder*

**Conclusions**

- Hyperacusis can have a major impact on quality of life.
- Proper referral to other specialists and accurate information are essential.
- Environmental sound enrichment and desensitization programs are recommended.
- AIT and other therapies should not be recommended due to lack of evidence based research.