### **Hyperacusis: Update on Diagnosis and Treatment**

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# HYPERACUSIS: 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)

# **HYPERACUSIS: Terminology and Definitions (2)**

- Hyperacusis
  - Abnormally strong reactions (intolerance) occurring within the *central* auditory pathways with exposure to moderate levels of sounds
  - Hearing sensitivity is typically normal
  - Otoacoustic emissions (OAEs) are typically normal
  - Lack of contralateral suppression of OAEs
  - Reduced LDLs
- Loudness recruitment
  - Abnormal growth of loudness
  - Sensory (outer hair cell) hearing loss (peripheral)
  - OAEs are abnormal
  - Reduced LDLs

# HYPERACUSIS: Hyperacusis versus Phonophobia

- Hyperacusis
  - General intolerance to any "loud" sound
  - Occurs with sounds of any frequency
  - Related to inadequate efferent system
- Phonophobia
  - Fear of sound
  - Anticipatory, anxious, and sometimes different reaction to specific sounds, e.g.,
    - ✓ Vacuum cleaner
    - ✓ Siren
    - **✓** Telephone ringing
    - ✓ Hair dryer
    - Crying child
    - ✓ Barking do
  - Reaction and avoidance to sounds involves learning and conditioning
  - Consistently related to intensity of sound
  - Intact peripheral and efferent auditory systems
  - Enhanced connections between the auditory cortical regions and the limbic and autonomic nervous system regions

# **HYPERACUSIS: Terminology and Definitions (3)**

- Misophonia
  - Dislike or hatred of sound (may be selected sounds)
- **□** Selective Sound Sensitivity Syndrome (SSSS or 4S)
  - Irritation and dislike of specific soft sounds
  - Most common sounds, produced by family members, e.g.,
    - ✓ Eating
    - ✓ Smacking lips
    - ✓ Breathing
    - ✓ Speech sounds

#### **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.

### HYPERACUSIS: Selected General References (1)

- □ Perlman H. Hyperacusis. *Ann Otol Rhinol Laryngol 47*: 1938.
- Marriage J & Barnes NM. Is central hyperacusis a symptom of 5hydroxytrypamine (5-HT) dysfunction? J Laryngol Otol 109: 1995.
- □ Jastreboff P, Gray WC, Gold SL. Neurophysiological approach to tinnitus patients. *Am J Otol 17*: 1996.
- □ Jastreboff PJ & Jastreboff MM. Tinnitus retraining therapy for patients with tinnitus and decreased sound tolerance. *Otolaryngol Clin North Am 36*: 2003. Phillips DP & Carr MM. Disturbances of loudness perception. *JAAA 9*: 1998.
- □ Andersson et al. Hypersensitivity to sound (hyperacusis): a prevalence study conducted via the Internet and post. Int J Audiol 41: 2002. [point prevalence = 6 7%]
- Katzenell & Segal S. Hyperacusis: Review and clinical guidelines. *Otol & Neurotol 22*: 2001.
- Baguley DM. Hyperacusis. *J Royal Society Med 96*: 2003.

# HYPERACUSIS: Book

Baguley David M & Andersson Gerhard. *Hyperacusis: Mechanisms, Diagnosis, and Therapies*. Plural Publishing: San Diego, 2007





### HYPERACUSIS: Selected Pediatric References (2)

- □ Johnson LB, Comeau M, Clarke KD. Hyperacusis in William's syndrome. *J Otolaryngol 30*: 2001.
- ☐ Gethelf et al. Hyperacusis in Williams syndrome. *Neurology 66*: 2006.
- □ Klein et al. Hyperacusis and otitis media in individuals with Williams syndrome. JSHD 55: 1990.
- □ Blomberg et al. Fears, hyperacusis and musicality in Williams syndrome. Research in Developmental Disabilities 27: 2006.
- Rosenhall et al. Autism and hearing loss. *J Autism Dev Disord 29*: 1999.
- □ Gopal et al. Effects of selective serotonin reuptake inhibitors on auditory processing: a case study. *JAAA 11*: 2000.
- □ Khalfa et al. Increased perception of loudness in autism. *Hearing Research 198*: 2004.

### HYPERACUSIS: Selected Adult References (3)

- Weber et al. Central hyperacusis with phonophobia in multiple sclerosis. *Multiple Sclerosis 8*: 505-509, 2002. [abnormal ABR findings in 3 patients; *hyperacusis first symptom*]
- □ Hallberg et al. Daily living with hyperacusis due to head injury 1 year after a treatment programme at the hearing clinic. Scand J Caring 19: 410-418, 2005.

# HYPERACUSIS: Websites

- Hyperacusis.org
  - Marsha Johnson
  - Oregon Tinnitus and Hyperacusis Clinic
  - Coined term 4S
- Hyperacusis.net
  - Internet support group
  - Maintained by laypersons (not audiologists)
  - Hyperacusis described as a disease or affliction
  - Includes some inaccurate information
- Hyperacusis.com

### HYPERACUSIS: A symptom in 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
- Pyrodoxine deficiency
- Benzodiazepine dependency
- William's syndrome
- Autism

### **HYPERACUSIS:** Williams Syndrome

- 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)

### HYPERACUSIS: A symptom of varied clinical entities (2)

- Tinnitus
- Acoustic trauma
- Auto-immune disorders
- Post otologic surgery
  - ventilation tubes
  - otosclerosis
  - tympanoplasty
- ☐ Genetic predisposition (family trait)
- Auditory processing disorders (APD)
- Drugs
  - Effexor
  - Prozac
  - Remeron
  - Tegretol
  - Zoloft

# Tinnitus and Hyperacusis in Childhood: Preliminary Findings in An APD Population at the University of Florida (N = 77)

	Yes	No
Tinnitus	10 (13%)	67 (87%)
Hyperacusis	30 (39%)	47 (61%)
<b>Both Tinnitus &amp; Hyperacusis</b>	8 (10%)	69 (90%)

## HYPERACUSIS: Prevalence in General Population

- Marriage & Barnes (1995): Prevalence unknown, but probably underestimated.
- □ Rubinstein et al (1996): 22% (no definition given)
- □ Rabijanska et al (1999): 15.2 (unclear methodology)
- Andersson, Lindvall, Hursti & Carlbring (2002):
  - Prevalence of 8% (postal survey)
  - Prevalence of 9% (internet survey)
  - Severe hyperacusis estimated in 2 to 3%

# 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
- Rabijanska et al (1999): 15.2 (unclear methodology)
- Andersson, Lindvall, Hursti & Carlbring (2002):
  - Prevalence of 8% (postal survey)
  - Prevalence of 9% (internet survey)
  - Severe hyperacusis estimated in 2 to 3%

### **HYPERACUSIS: In Tinnitus Population**

- Bartnik et al (1999): 40%
- □ Hall (1999): 54%
- □ Jastreboff & Jastreboff (2000): 40%
- Andersson et al (2001): 60%
- □ Dauman & Bouscau-Faure (2005): 79%

# HYPERACUSIS: Examples of sounds considered aversive by persons with hyperacusis (N = 1151) (Andersson et al, 2002)

#### What kind of sound to you consider aversive?

	%	N
Noise	57	660
Music	27	309
Talk	3	39
Paper noises	5	55
Clatter	15	171
Mechanical	28	326
Other everyday sounds	24	274

### HYPERACUSIS: Sounds that are most bothersome (various sources plus University of Florida clinic)

- Shrill sounds
- Power saw
- **□** Telephone ringing
- Vacuum cleaner
- Hair dryer
- ☐ Sirens (e.g., ambulance)
- □ Children crying and screaming

# HYPERACUSIS: Reactions when being exposed to annoying sounds (N = 1157) (Andersson et al, 2002)

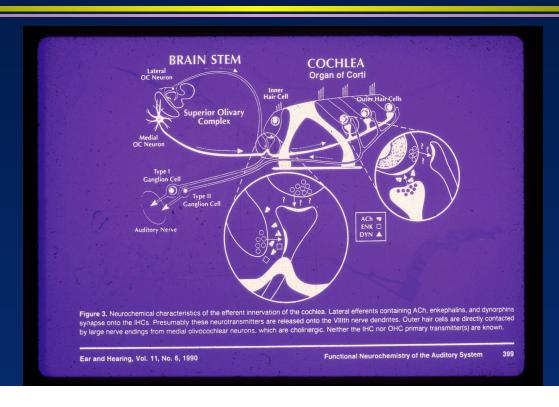
How do you feel when you are being exposed to disturbing sounds?

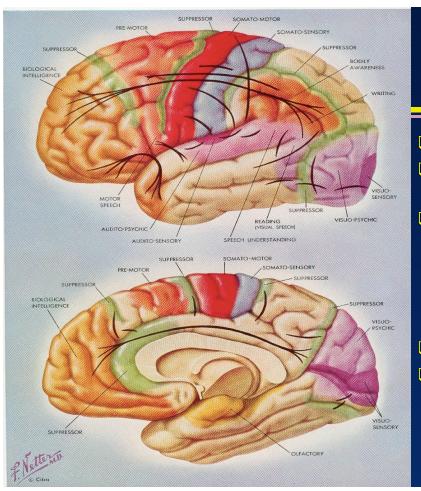
	<b>%</b>	N
Tense	10	119
Angry	12	141
Irritated	75	862
Afraid	1	16
Poor concentration	41	479
In pain	5	57

### HYPERACUSIS: Possible mechanisms

- Imbalance of neuro-chemical seratonin (5 HT), involved in
  - stimulus reactivity
  - perception of sensory information in brain
- Release of neuro-transmitter 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: Possible biochemical mechanisms involving efferent auditory system





### **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: ASSESSMENT AND MANAGEMENT CONSULTATION AUDIOLOGIC ASSESSMENT REFERRAL FOR AUDIOLOGIC EVALUATION OR Rx MANAGEMENT**

#### **Tinnitus Handicap Inventory**

(Newman, Jacobson & Spitzer.

Arch Otolaryngol Head & Neck Surg 122: 1996)

- 25 items (can use to assess impact of hyperacusis on quality of life)
  - 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?"

### **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
    - ✓ Pain
    - ✓ Annoyance
    - ✓ Uncomfortable
    - ✓ 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

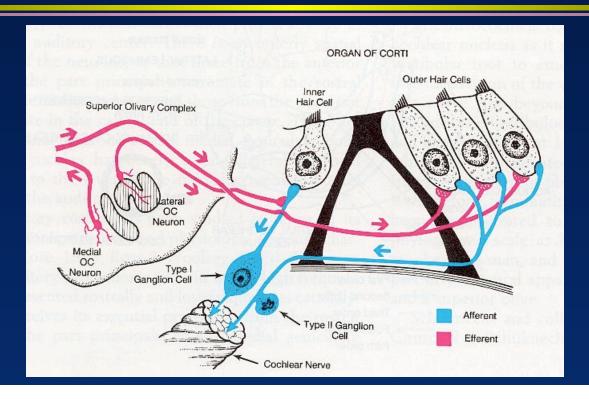
#### **HYPERACUSIS ASSESSMENT**

- Immittance measurement (no acoustic reflexes for patients with hyperacusis)
- □ Distortion product otoacoustic emissions (DPOAE) for 500 to 8000 Hz ( 6 frequencies/octave)
  - Suppression of OAEs with ipsilateral & contralateral noise
- Pure tone audiometry
  - Inter-octaves (3000 and 6000 Hz)
  - High frequency audiometry to 20,000 Hz
- Word recognition scores (most comfortable level)
- Measure loudness discomfort levels (LDLs) for tones and speech sounds (to identify hyperacusis)
- Neuro-diagnostic auditory brainstem response (ABR) as indicated (patient refuses MRI due to high noise levels)

#### **DPOAEs in HYPERACUSIS ASSESSMENT**

- □ Sztuka A, Pośpiech L, Gawron W, Dudek K (2006)
  - Subjects were patients with tinnitus, including subgroup also with "hyperacusis and misophonia"
  - "Hyperacusis has important influence on DPOAE amplitude; essentially increases amplitude of DPOAE in the examined group of tinnitus patients."

# **HYPERACUSIS ASSESSMENT:**Contralateral suppression of OAEs



## **HYPERACUSIS:** Consultation (1)

#### ■ 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?
- Audiologic history, e.g., infant risk indicators, previous audiograms, CAPD, tinnitus?
- Other related disorders, e.g., sensory integration disorder, autism spectrum disorders?

# **HYPERACUSIS:** Consultation (2)

- 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
  - avoidance of noisy places (alteration of daily activities)
  - running away or potentially dangerous evasive actions

# **HYPERACUSIS:** Consultation (3)

- Attempt to answer all parent/child questions
- □ Definition 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

### **Patient/Family Counseling and Education**

"Knowledge is power."

(Nam et ipsa scientia potestas est.)

**Francis Bacon** (1561-1626)

Meditationes Sacrae [1597]

### **HYPERACUSIS: Audiologic Management**

- Desensitization
  - Suggestions for home management, e.g., honest discussions about bothersome sounds
  - Tape record sounds and replay 10 minutes/day louder each day
  - Avoid silence (no earplugs unless indicated by behavior and/or noise levels)
- **■** Extended management options
  - Retraining therapy (TRT)
    - **✓** Directive counseling
    - ✓ Noise generator fitting
  - Neuromonics Tinnitus Treatment
- Referral to other professionals
  - Occupational therapist
  - Neurologist
  - Otolaryngologist
  - Psychologist/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
- 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)

### HYPERACUSIS: Evidence in Support of Management with TRT

- Formby et al (2008)
  - Randomized, double-blind, placebo-controlled clinical trial of efficacy of TRT in hyperacusis
  - 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
  - Over 80% of subjects assigned full treatment group achieved significant benefit, I.e.,
    - ✓ Increase in > 10 dB in LDLs and Contour Test for Loudness
    - ✓ Over 80% of subjects assigned full treatment group achieved significant benefit, i.e.,Increase in > 10 dB in LDLs and Contour Test for Loudness
  - Most subjects assigned to partial treatment group did not benefit from treatment

### HYPERACUSIS: Evidence in Support of Management with TRT

- Herraiz, Plaza & Aparicio (2006) Spain
  - Review of hyperacusis management with TRT
- 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
- Noreña AJ, Chery-Croze S (2007) France
  - 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

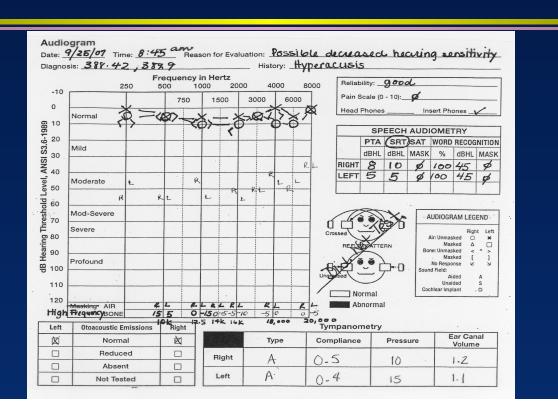
# Neuromonics Treatment for Hyperacusis: Five Step Treatment (Promising but no published evidence)



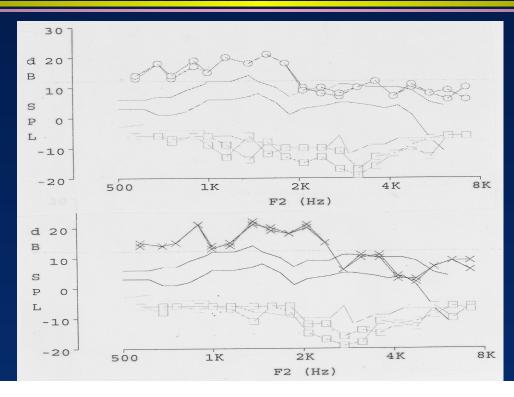
### HYPERACUSIS: What about Auditory Integration Theory Management?

- Sound therapy program introduced by French physician Guy Berard ("retrained from a surgeon to a hearing specialist" now retired)
  - "Dr. Berard explained that, if we brought Georgie in to see him, he would do a
    detailed audiogram which would reveal as accurately as possible the exact
    frequencies where her distortions occurred." (The Sound of a Miracle", p.
    157)
- □ Classified as "experimental" by AAA and ASHA
  - No independent clinical trials (double-blind with control group)
  - No formal assessment of sound levels for sounds used in treatment
  - Important "placebo effect" with treatment
  - Illiogical theoretical assumptions, e.g.,
    - ✓ Training muscles in middle ear
    - ✓ Training hair cells in inner ear

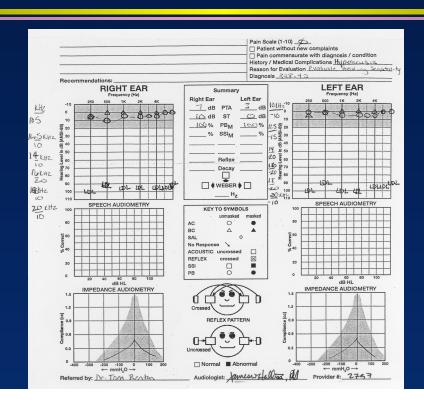
#### **Adult with hyperacusis: Audiogram and LDLs**



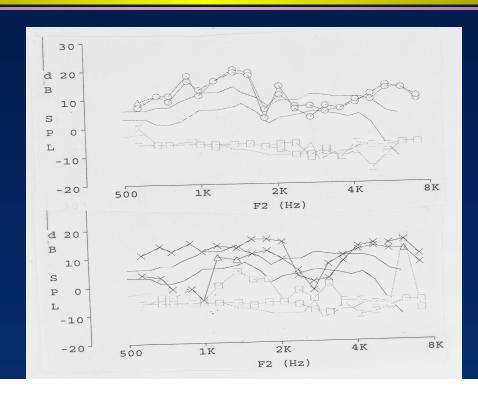
## Adult with hyperacusis: Distortion product otoacoustic emissions



#### **Child with hyperacusis: Audiogram and LDLs**



## Child with hyperacusis: Distortion product otoacoustic emissions

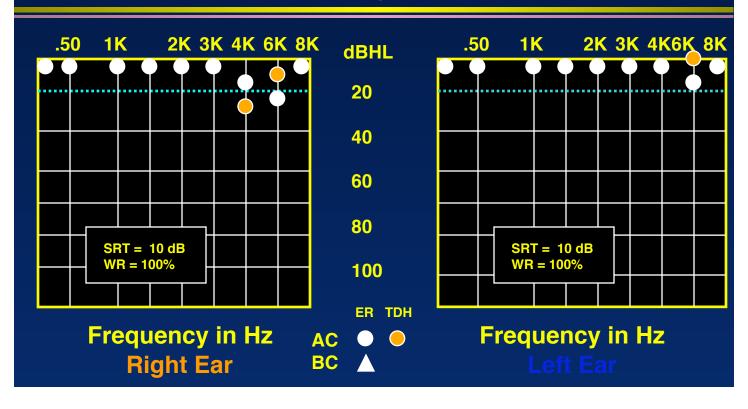


- 18 year old female
- ☐ Freshman at the University of Florida (music major)
- □ Referred by out of town otolaryngologist
- History
  - Onset 10 months earlier when reportedly "damaged ears in loud recording studio"
  - Hyperacusis is worse in morning
  - Cannot tolerate everyday environmental sounds
    - ✓ 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

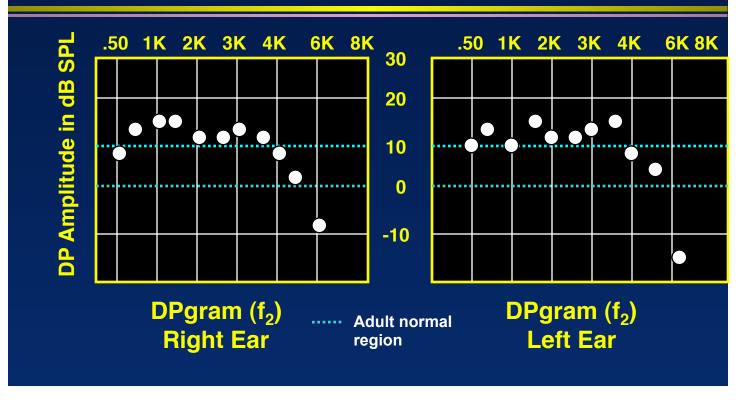
#### CASE REPORT: Therapy for Hyperacusis in Young Adult Audiogram



#### **High Frequency Audiometry**

Frequency (Hz)	Right Ear	Left Ear	
10,000	0	0	
12,500	0	0	
14,000	0	0	
16,000	0	0	
18,000	0	0	
20,000	0	0	

 $(L_1 = 65 \text{ dB SPL}; L_2 = 55 \text{ dB SPL}; f_2/f_1 = 1.2; 5 \text{ freq/octave})$ 



#### **Loudness Discomfort Levels (LDLs)**

Frequency (Hz)	Right Ear		Left Ear	
	PT	NBN	PT	NBN
1000	70	65	70	60
2000	75	60	75	60
4000	80	75	80	75
6000	75	65	75	70
8000	70	65	80	65
Speech	75		65	

### CASE REPORT: Hyperacusis in Young Adult 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

### CASE REPORT: Hyperacusis in Young Adult 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

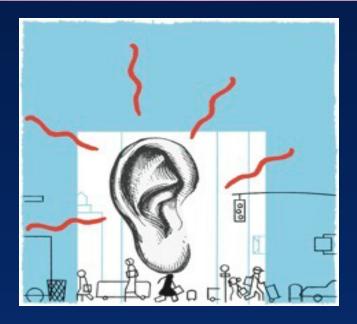
### Loudness Discomfort Levels (LDLs) for Pure Tone Signals

Frequency (Hz) Right Ear	Left Ear
Initial 1 Year	Initial 1 Year
1000 70 95	70 95
2000 75 95	75 90
4000 80 100	80 90
8000 70 > 90	80 > 90

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

- 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 counseling 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

## Thank you! *Questions?*



From: Groopman J. (February 29, 2009). That buzzing sound. The New Yorker.