Otology seminar

Hyperacusis: review and clinical guideline

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Definition:
- Mathisen 1969: Hyperacusis: increased sensitivity to sound in levels that would not trouble a normal individual
- Painful or troublesome sensation
- Phonophobia, noise sensitivity, audiosensitivity, reduced tolerance to noise, dysacusis, hyper-recruitment, misophonia
- Machinery sound, quiet sound, Tiredness and anxiety aggravated symptoms

Hyperacusis vs. recruitment
- Separate phenomenon
- Moore 1985:
  - Recruitment: associated with SNHL, cochlear outer hair cell damage
  - Narrowing the dynamic range caused by higher detection threshold
  - LDL unaffected
  - Damage to the outer hair cell, separated phenomenon, Noise-induced hearing loss
- Tzaneva 1996: Reduced LDL, 120 → 70 dB, narrowest dynamic range at 4000Hz with hearing loss > 20 dB.
  - Recruitment with malfunction of central regulation
- Central hyperacusis: normal audiogram
- Relationship? Not clear

Central hyperacusis vs. peripheral hyperacusis
- Audiogram: normal
- Recruitment usually negative

Primary and secondary central hyperacusis
- Primary: 5-HT function
- Secondary: learned association of discomfort with exposure to sound
  - “Behavior modification”
**Etiology:**
1. Peripheral auditory system
2. Nervous system
3. Hormonal
4. Infections

**Peripheral auditory system**
- Bell’s palsy
  - Adour 1974: 29% Bell’s palsy with hyperacusis.
  - McCandless 1974: Lower LDL (Loudness discomfort levels), “rollover” sign, some absence stapedial function
  - Collard 1984: Reduced amplitude of ipsilateral ART, Loss of CN VIII downregulation of the stimulus: mediated acoustic tensor reflex

- Stapedectomy
  - McCandless 1974: no reduction in Pure-tone LDL, but speech stimulation LDL decreased.
  - Operated ear: Reduction 7.7 dB. ABLB (alternate binaural loudness balance) change (+)
  - “Rollover” at 80-90 dB, changes of resonance.

- Ramsay Hunt syndrome
  - Polycranial neuritis
  - Wayman 1990: 46% hyperacusis in speech frequency, 31% in high-frequency hearing loss

  - Fukaya 1988: idiopathic perilymphatic fistula 62.5%
  - Gordon 1988: change in perilymphatic pressure

  - Axelsson 1987: 13% acoustic trauma
  - Gordon 1991: Meniere’s disease

**Central nervous system**
- Headache
  - Vingen 1998, Main 1997: >70 % Migrane → phonophobia, 83% during attacks, and 76% between attacks
  - Vingen 1998: Cluster headache: LDL difference 12.1 dB, unilateral or pulsating
Concomitant sensitivity to light and other sensory stimuli suggests a central processing mechanism
Main 1997: derangement in the serotoninergic midbrain raphe nuclei

**Depression**
- Carman 1973: associated with depression
  - Rodent: depletion of brain serotonin increased the magnitude of auditory startle response
  - Imipramine: better effect on depression patient with hyperacusis
- Gordon 1981: Benzodiazepine withdrawal \(\rightarrow\) lower threshold of ART and intolerance to noise

**Minor head injury**
- Waddell 1984: Lower LDL: 82/94 dB
- No correlation of LDL with hyperacusis

**Williams’s syndrome**
- Nigam 1994: rare genetic disorder
  - Involve vascular, connective and CNS
  - Deletion on Chromosome 7, elastin gene locus
  - Sporadic inheritance
  - Diagnosis: 3/5, Facial appearance (elfin face), cardiovascular abnormalities, growth retardation, developmental retardation, infantile hypercalcemia
  - 95% hyperacusis
  - Absence of TEOAEs with normal hearing: lack of central modulation and loss of OHC inhibition

**Learning disability and stuttering**
- MacCulloch 1971: LDL of stutterers was lower
- Gorgon 1986: 93 patient with abnormal acoustic reflex
- Associated with autism and developmental disorder

**Tinnitus**
- Jastreboff 1993: Hyperacusis often occurs with tinnitus before its onset
- Increased central gain, pretinnitus state, for early detection
• Spinal problems
  ■ Gordon 1991: Hyperacusis after spinal anesthesia
    ◆ Fluctuation of CSF pressure within the cochlea
  ■ Oen 1997: Hyperacusis in young children with spinal bufida

  ■ Cohen 1988: Multiple sclerosis of left olivocochlear bundle in the pons, lower LDL, lower ART 5-10 dB, Efferent fibers from this bundle reach the cochlea bilaterally and produce an inhibitory effect

  ■ Fukutake 1998: TIA, lesion at right posteroinferior thalamus and medial geniculate body, pontine auditory hallucinosis.
  ■ Temporal lobe lesion also been reported

**Hormonal and infectious diseases**

• Addison’s disease
  ■ Hekin 1968: dynamic auditory range is significantly decreased in patients with Addison’s disease and panhypopituitarism
    ◆ Correcting effect of steroid replacement therapy: absence of carbohydrate-active steroid increase detection sensitivity and decreases auditory perceptual and integration abilities.
    ◆ NCV of peripheral and central nerve changes
  ■ Brandy 1995: Hyperthyroidism and hyperacusis

• Lyme disease
    ◆ Treatment with carbamazepine diminished the hyperacusis
    ◆ Involve the Limbic system and the amygdala, psychophysiologic effects through GABA receptor
  ■ Perlman 1938: Neurosyphilis with hyperacusis

**Pathophysiology of hyperacusis**

• The 5-hydroxytryptamine hypotheses
  ■ Davis 1980: Startle response of rats, serotonin inhibits auditory input in the forebrain
  ■ Davis 1986: stimulus reactivity and sensory reception, activation of 5-HT1B receptors in the forebrain appears to depress the startle reflex
  ■ Maesden 1989: anxiety control
Lance 1991: Migrane headache, 5-HT\textsubscript{1D} and 5-HT\textsubscript{2C}
Biler 1994: altered in depression
Marriage 1995: malfunction of 5-HT

TABLE II

<table>
<thead>
<tr>
<th>Receptor subtype</th>
<th>Operational characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>5-HT\textsubscript{1A}</td>
<td>Hypotension, anxiety, depression</td>
</tr>
<tr>
<td>5-HT\textsubscript{1B}</td>
<td>Inhibition of neurotransmitter release, startle and sensory modulation (Davies et al., 1986)</td>
</tr>
<tr>
<td>5-HT\textsubscript{1D}</td>
<td>Inhibition of neurotransmitter release, migraine</td>
</tr>
<tr>
<td>5-HT\textsubscript{2A}</td>
<td>Vasoconstriction, platelet aggregation, depression</td>
</tr>
<tr>
<td>5-HT\textsubscript{2B}</td>
<td>Rat stomach fundic muscle contraction</td>
</tr>
<tr>
<td>5-HT\textsubscript{2C}</td>
<td>Migraine, anxiety and eating disorders</td>
</tr>
<tr>
<td>5-HT\textsubscript{3}</td>
<td>Emesis, anxiety and psychosis</td>
</tr>
<tr>
<td>5-HT\textsubscript{4}</td>
<td>Gastric stasis, tachycardia</td>
</tr>
</tbody>
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- Role of 5-HT
  - Brewerton 1988: 5-HT agonists induce migraine headaches
  - Kennett 1989: Benzodiazepine reduces 5-HT concentration, withdrawal \(\rightarrow\) tinnitus and hyperacusis
  - Andron 1989: 5-HT and generation of auditory hallucinations
  - Meltzer 1991: 5-HT receptor in treatment of chronic depression
  - Wilson 1992: 5-HT and genesis of fatigue in man

- The neuroactive opioid peptides hypothesis
  - Sahley 1996: Descending axon arises form lateral efferent brainstem nuclei \(\rightarrow\) auditory type I spiral ganglion dendrites \(\rightarrow\) cochlear inner hair cells
    - Within ipsilateral cochlea
  - Ach + opioid \(\rightarrow\) increase auditory sensitivity and ABR
  - For heightened state of auditory vigilance during actual or perceived life-threatening situations

- The plasticity of the ascending auditory system hypothesis
  - Niemeyer 1971: industrial noise \(\rightarrow\) LDL rises
- Central adaptation when cochlear sensitivity constant
  - Szczepniak 1996: Noise exposure $\rightarrow$ decrease inhibition in the cochlear nucleus $\rightarrow$ increase the acoustic evoked activity in inferior colliculus (IC) $\rightarrow$ increased IC wave
  - Decrease GABAergic inhibition on IC neurons
  - Tone exposure decreases GABA-mediated inhibition on temporal integration in the IC
  - Baclofen

- The cochlear hypothesis
  - Axelsson and Anari 1993: Hyperacusis + tinnitus + distortion $\rightarrow$ cochlear origin
  - Perilympathic fistula, after stapedectomy, Bell’s palsy, Ramsay Hunt syndrome, Meniere’s disease and SNHL

Suggested clinical guideline

- Healthy history
  - What kind of noises and what length?
  - Bilateral or unilateral?
  - Otologic disease? Otologic surgical procedure?
  - Hearing loss? Noise exposure or acoustic trauma?
  - Associated tinnitus or vestibular problems? Any facial symptoms?
  - Headache? Aura? Parethesesas?
  - Muscle cramps?
  - Fatigue? Loss of appetite? BW loss? Skin or mucosal discoloration? GI symptoms?
  - Depression?
  - Cranial trauma? Drug abuse?

- Physical examination
  - Facial paralysis? Skin or mucosal discoloration? Auricular lesion? T/P/R?
  - Neurological examination of Cranial nerve: motor, sensory, reflex and fundoscopic examination

FIG. 1. A block diagram of the stages of tinnitus development and the main centers of the nervous system involved in the emergence of tinnitus perception and in determining its annoyance level. Functional interrelations are described in the text.
• Laboratory
  ■ CBC, electrolyte, cortisol, TSH

• Audiologic test
  ■ PTA, SRT, LDL, ABLB, ART, OAE
  ■ Brandy 1995: peripheral lesion $\rightarrow$ ART difference
  ■ Thornton 1987: define LDL with ABR
    ◆ LDL=$I + 15 \text{ dB}$
    ◆ $I=$intensity in which the wave V latency/intensity slope $< 0.1\text{msec}/10\text{dB}$

**Treatment:**
**Treat underlying first**
**Reassurance!**
**Avoid silence!**
◆ With hearing loss $\rightarrow$ Hearing aid usage!
◆ Without hearing loss:
  ■ Earplugs or protective devices
  ■ Auditory integration training (AIT): auditory enhancement training
  ■ **Tinnitus masker:** white nose, hyperacusic desensitization or successive approximation
  ◆ Hazell 1991: 27/30 successes, LDL increase 5-10 dB
  ◆ Jastreboff 1996: average 14 dB, as much as 30 dB in 18 months
    ● Counseling alone 18%
    ● Hearing aids: 70%
    ● Noise generator: 83%
  ■ **Electronic attenuator**
    ◆ Binaural Micro-Tech
      Refuge-hyperacusic instruments
      ● 4:1 compression ratio
      ● 90 dB $\rightarrow$ 70 dB SPL
    ◆ Cherry 1996: cochlear labyrinthectomy for profound hyperacusis

![Signal](image)

**FIG. 2.** Relation of tinnitus signal to neuronal activity under different conditions. Tinnitus under normal conditions (upper panel); masking of tinnitus by high level of the noise (middle panel); interference with tinnitus signal by low-level noise (lower panel).
Reference:


