



Experience of Hyperacusis in Children

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What is Hyperacusis?

- Intolerance and aversion of sounds that would be tolerated by most listeners
 - Avoidance of situations where sounds may occur (phonophobia)
 - “Hyperacusis” infers a cochlear pathology but no evidence that there is increased sensitivity to threshold detection levels.
 - Not the same as recruitment
 - Not demonstrated as peripheral pathology
- THOUGH MAY CO-OCCUR with HEARING LOSS**

How can we test for hyperacusis?

- No objective test for hyperacusis
- Subjective condition, need validated questionnaire
- Children may not be able to describe sensations
- Behavioural symptom profile (crying, cowering, covering ears, lashing out, running away)
- Parent description of behaviours
- Can develop into “phonophobia” with very similar behavioural profile

So, how do we know it has a physiological basis?

Prevalence of loudness sensitivity in typical school-age population

Coelho, Sanchez and Tyler (2007)

N= 506 typically developing school children

From 5 to 12 years in 15 schools

- Measured LDLs “loudness sensitivity” if below 5 percentile = <90 dBHL
- “Loudness annoyance” if “bothered by sound” and list of 10 or more /20 sound sources (42%)

If both, considered to have “hyperacusis”

Prevalence of Hyperacusis: 3.2%

of whom 50% had tinnitus, phonophobia in 9% (scared by sound)

Risk factor was mild hearing loss in left ear.

Two lectures from Day 1: Kevin Munro: FORMBY et al (JASA, 2003,114, 55-58)

Measured loudness rating in 10 normal-hearing adults pre & post treatment.

Two sound treatments:

earplugs (attenuation) => loudness rating higher

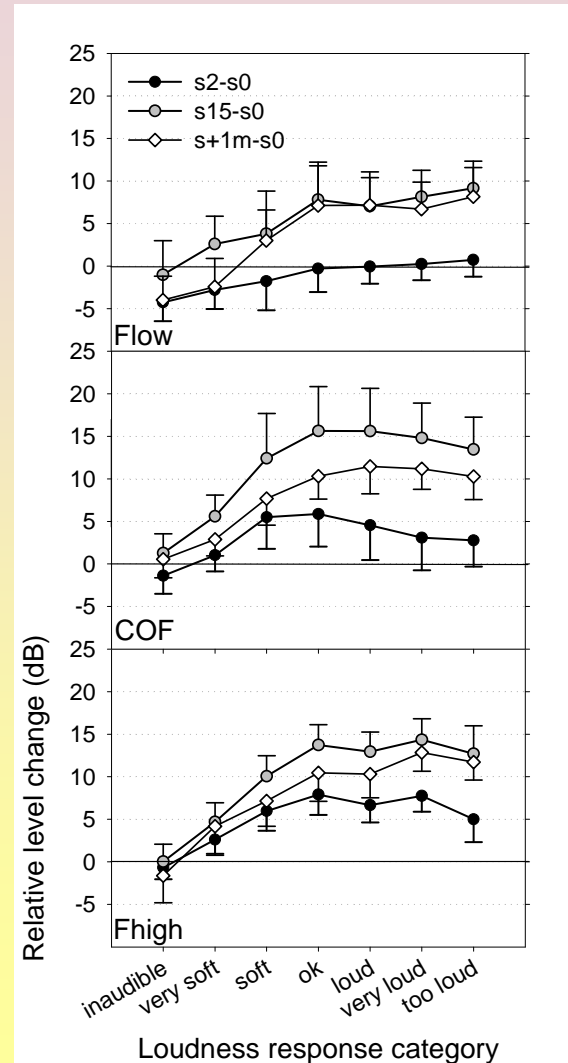
broadband noise generators (enrichment) loud rating lower

Munro et al used ARTs

1. clear demonstration of experience-related changes in sensory processing in adult auditory brainstem
2. results consistent with a change in central gain
3. findings suggest that using earplugs will not help individuals with hyperacusis

Arnaud Norena Day 1: Acoustic enhancement Norena & Chery-Croze (2007)

Acoustic enhancement improves hyperacusis



General Hyperacusis / Tinnitus literature

Probably physiological basis to hyperacusis

Studies show threshold for ART reduces after ear plugs (Formby et al, Munro et al, 2009)

Maintained by fear/anxiety (amygdala)

Enhanced auditory vigilance

Needs effective management of all aspects

Acoustic enhancement reduces anxiety *and* auditory sensitivity. Tyler reports 2/3 children benefit

Plugging of ears *increases* auditory sensitivity. Do not use ear plugs for hyperacusis

Known paediatric populations with susceptibility to sound intolerance

- Autistic spectrum disorders (ASD) 17-20%
- Williams Syndrome (90%, from early life, anxiety)

Unknown prevalence:

- Auditory Processing Disorders (APD)
- Migraine
- Attention Deficit Disorder (ADD)
- Head Injury / Hydrocephalus
- Tinnitus without hearing loss

Research literature: Untreated hearing loss exacerbates auditory sensitivity

- Mild, open-fit amplification very successful for improving hyperacusis,
- Hyperacusis occurs with CHL, SNHL (WS also)
- Implies abnormality in central auditory processing
- No association with measured level of sound source

Indicates: Hearing Loss needs proactive management and amplification.

Hyperacusis is not a contra-indication for hearing aids or surgery

Anxiety or Heightened Sensitivity or both?

Gallo et al study: 2007 (n=21 WS)

- 90% WS showed overt behavioural reactivity to mild intensity sounds cf 20% in dev. delay control group
- Video analysis shows behaviours BEFORE onset of sound, inferring anticipatory anxiety
- Greater acoustic startle eye blinks: heightened emotional state. Good quality research, needs further assess.

IMPLIES: cannot separate fear component, becomes established *with* auditory sensitivity.

Control group for developmental delay, not specific sensory-profile groups

Management of fear/anxiety and auditory aspects



Video of subject with permanent conductive hearing loss (BOF synd)

Branchial Ocular Facial Syndrome (dominant aut rec)

- Permanent conductive hearing loss, cleft palate
 - Visual impairment
 - Learning disability
 - Daniel has addition diagnosis of Autistic Spec Dis
 - Hearing aids from 7 years, improved school prog+
- Hyperacusis Major Management issue.

Same severity With and Without Hearing Aids

Audiogram

Unaided

Sp disc %

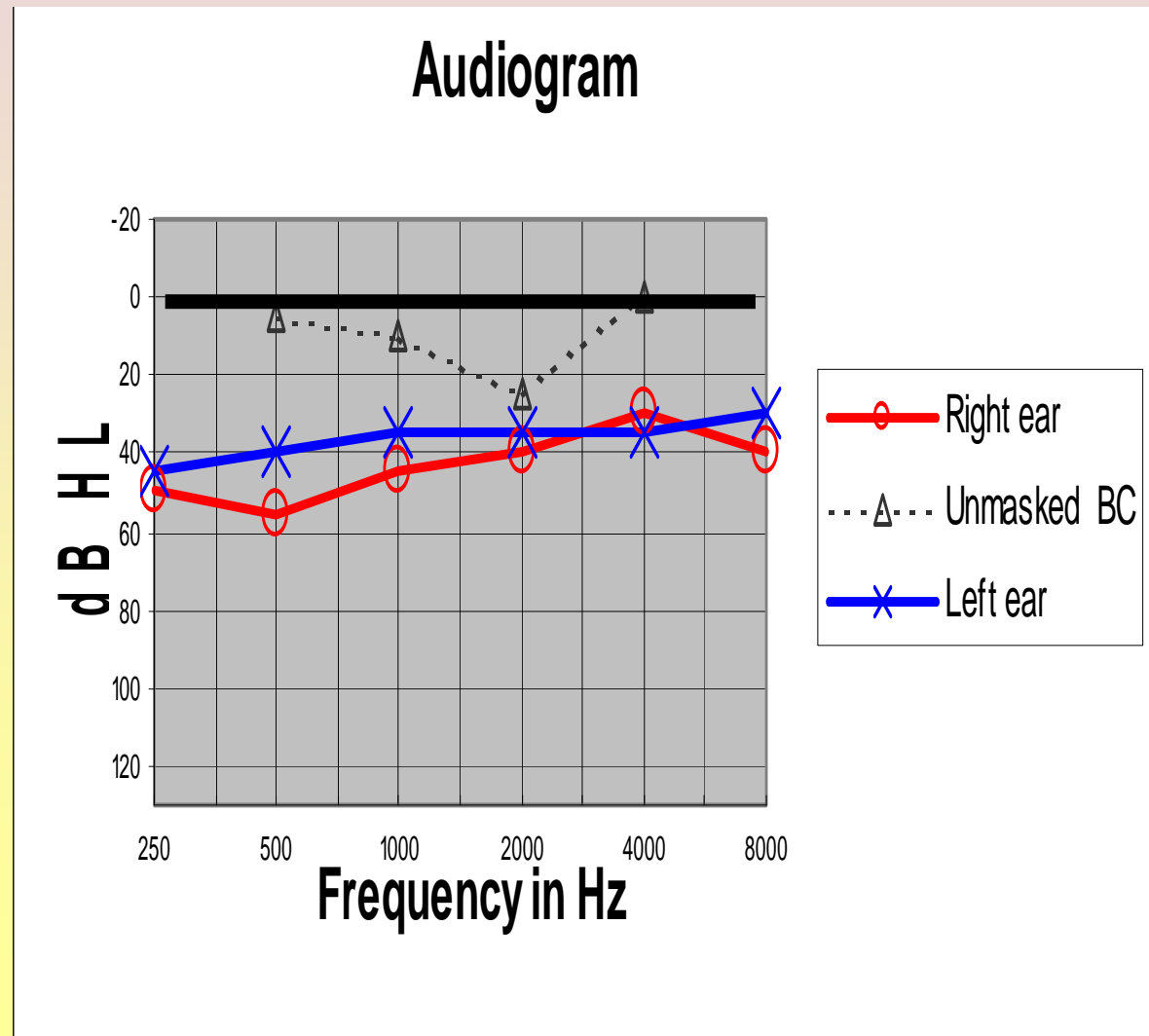
70 dB 94

60 dB 70

50 dB 10

With WNG

70 dB 66





Hyperacusis profile:

Sound aversion from young life “hurts ears”

Pain continues after noise stops.

Babies crying, dogs barking, balloons, party poppers

Becomes uncontrollable, unpredictable and rapid

Response: runs away into danger

Now 19 yrs “so sad, missing out on things he enjoys,
it is controlling our lives, we really need help

Extent of **problem on 0-10 SCALE** given as **20**



Aged 19 years, fitted with bilateral silent star White Noise Generators (WNG) on open moulds

- First day child screaming in supermarket, no concerns, no fear-potentiated sensitivity
- Wears “my little stars” on volume 4 when goes out
- As becomes settled turns down.
- Old and New situations, easy, able to retain control

PROBLEM: can't hear speech at all

Therefore needs hearing aids with WN option, closed moulds to prevent feedback (Active wave)



Video clips of Daniel and mother



Needs hearing aids and to listen to music

Evidence-base for improvement?

1. Case study: Effective use of HA all settings:
T setting for cinema, able to go to new situations
Converter to listen to music MP3 player
2. SCALE of extent of problem: 4 “anxiety improved”
3. Diary forms of limited help, Questionnaire?
4. Need to follow up over longer time course
SENSE very helpful (dual sensory impairment).



Is it anxiety management or hearing management?

- Primarily anxiety at this stage, but a hearing component also.
- With WNG: No pain on exposure to sound
- Over time and with HA use predict change in central plasticity from acoustic enhancement

Professionals need to keep an open mind to impact of hyperacusis

Cannot be diagnosed by audiogram
and ULLs/ARTs

Questions? Comments?

References:

- Coelho, Sanchez and Tyler (2007) Hyperacusis, sound annoyance and loudness hypersensitivity in children. *Prog Brain Res* 166: 169-78.
- Levitin et al (2002) Neural correlates of Auditory Perception in Williams Syndrome: An fMRI study. *NeuroImage* 18, 74-82.