Aural diversity, decreased sound tolerance and Audiology

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Aural diversity

Everyone hears differently
Hearing ability changes through the lifespan
- Maturation
- Presbycusis
Hearing loss (congenital, acquired)
Auditory processing function
Sound sensitivity/decreased sound tolerance

Aural Diversity project
www.auraldiversity.org
What is normal hearing?

Equity, equality, removing barriers
Aural (re)habilitation

Improve access to sound for the purpose of –
Communication
Safety
Enjoyment

Aural typical versus aural diverse

World is designed for “aural typicals”: those with healthy, “normal” auditory systems
- Acoustic regulations
- Public announcement systems
- Building design
- Headphones
- Entertainment
- Sound installations in the arts

Not everyone has the same listening capacity

Topics for today

- How do we define DST?
- What are the proposed mechanisms of DST?
- How do we best identify and measure DST?
- Treatment options and their efficacy?
- Barriers to sound access
- Future directions in research
Decreased sound tolerance
Negative reaction to sound that the average listener doesn't experience

When does sound become noise?
Noise is unwanted sound
- Harmful
- Impedes communication, sound detection (SNR)
- Too loud
- Evokes negative emotions (fear, anger, annoyance)
- Causes discomfort
- Signifies a possible threat (fear)
- Increases tinnitus

Context of sound
Reduced tolerance may vary with
- Expected/unexpected
- Sound source
- Ambient sound levels
- Mood
DST: Definitive definition

Needs to encompass the various reactions to sound that patients experience:
- Loudness
- Emotions:
  - Annoying
  - Distressing
  - Fear-inducing
  - Overwhelming
- Physical:
  - Pain, discomfort
- Cognitive:
  - Awareness
  - Attention

Decreased sound tolerance

An umbrella term that includes:
- Hyperacusis
- Misophonia

Hyperacusis

Consensus-agreed definition (UK)

"A reduced tolerance of or an increased sensitivity to sound(s) that are perceived as normal to the majority of the population, or were perceived as normal to the person before the onset of their hyperacusis."

Adams et al. in review
Jastreboff & Jastreboff (2014)
Decreased sound tolerance: Inability to tolerate everyday sounds that don't bother other people
- Hyperacusis: A negative reaction to sound based on its physical characteristics
- Misophonia: Abnormally strong reaction to sound with a specific pattern/meaning
- Phonophobia: Fear is the dominant emotional reaction

Misophonia
Negative emotional reaction to specific sounds
Also described as “selective sound sensitivity syndrome” or “4S”
Often develops in childhood, adolescence
Family history common

Misophonia “trigger sounds”
- Chewing, eating, crunching
- Lip smacking
- Pen clicking
- Clock ticking
- Low-frequency, bass sounds
- Footsteps
- Fingers tapping
- Whistling
- Keyboard clicking
- Plastic bags
- Repetitive barking
- Sniffing
- Edelstein et al. 2013
Reaction to trigger sounds
Emotional: irritation, disgust, anger, rage; poss. intense anxiety, panic (not fear)
Physical:
- pressure in the chest, arms, head, or entire body
- clenched, tightened, and tense muscles
- increase in blood pressure, heart rate or body temperature, sweaty palms
- physical pain
- difficulty breathing
Physical aggression may result
- Edelstein et al. 2013

One person’s story

Is misophonia distinct from other forms of DST?

Reaction to sound
- Enhanced emotionally, physically
- Hyper-attentive to sound
- PWM may recognised reaction as unreasonable
- Can have profound impact on ability to engage with sound, many ADLs
- May intensify with time

Sounds always made by others
82% made by particular individuals (Edelstein et al. 2013)
May have visual triggers, too
Mechanisms
Unrelated to auditory function
Not reported as result of trauma (although trauma may exacerbate mood etc.)
Psychiatric (Schroeder et al. 2013):
• Shares features with many psychiatric conditions (e.g. OCD)
• Distinct psychiatric condition? Anxiety?
• Amsterdam Misophonia Scale
Neurologic (Edelstein et al. 2013):
• Similar neurologic basis as synesthesia? (sandpaper → jealousy)

Phonophobia
Fear of sound
• Damage
• Sensations
• Tinnitus alternations
Also used to describe
• Noise sensitivity in migraines
• Reduced sound tolerance in neurology

Tyler et al. (2014)
Loudness hyperacusis
Annoyance hyperacusis
Fear hyperacusis
Pain hyperacusis

Loudness hyperacusis

Moderately intense sounds are judged to be very loud compared to what a normal hearing person would experience.

Distinct from loudness recruitment

Loudness recruitment (LR)

- Abnormal loudness growth
- Cochlear phenomenon
- Does not vary with mood
- Not a treatable condition
- Clinical implications
  - Lowered sensation levels for acoustic reflex testing
  - Reduced dynamic range as thresholds “head south”

Loudness hyperacusis
Decreased sound tolerance of low/moderately loud sounds
May develop irrespective of hearing loss
May increase with stress, mood changes
Can have loudness recruitment *and hyperacusis

Comorbidities / associations
Peripheral
- Bells palsy
- Ramsay Hunt syndrome
- Post-stapedectomy
- Third window

Central
- Tinnitus
- Migraine
- Anxiety/depression
- PTSD
- Head injury
- Lyme disease
- Autism
- Williams syndrome
- Dementia
- Multiple sclerosis
Birds + hyperacusis

Loudness perception as measured by reaction time (RT)
RTs are shorter at moderate and high sound levels in HF SNHL canaries than NH canaries
Reduced RT suggests altered loudness perception/hyperacusis
- Lauer & Dooling, 2007

Relation with tinnitus

Patients with tinnitus
- 40% have hyperacusis
Patients with hyperacusis
- 86% have tinnitus

Proposed mechanisms

Loss of balance between exception and inhibition due to
- Dysfunction of the efferent system (both medial and lateral systems have been suggested)
- Dynorphins (endogenous opiates) increased as stress response. Enhances glutamate.
- 5-HT (serotonin) inhibitory regulator of central sensory processing
- Disruption could result in central hyperacusis (Marriage & Barnes, 1995)
- Ventral cochlear nucleus - changes to bushy cells after noise exposure
HYPERACUSIS
Abnormal enhancement of sound evoked neural activity in auditory pathways
Secondary activation of the limbic system and ANS

MISOPHONIA
Normal sound evoked neural activity in auditory pathways
Abnormal exaggerated activation of the limbic system and ANS

Mechanisms:
Enhanced bone conduction
Superior canal dehiscence (third window)
Loud sounds/pressure change → vertigo, “feel off” for duration of sound
Pulse-synchronous tinnitus
Fogginess, non-specific dizziness or light-headedness
Somatosounds:
- Autophony, better lying down (as with PET)
- Own footsteps
- Neck vertebrae clicking
Diagnosis:
- Air-bone gap with normal immittance
- VEMP’s (lower threshold in SCD)
- CT scan (bilateral in 10-50% of cases)
- 128 Hz tuning fork on ankle heard in ear

Pain hyperacusis

*“One of my sensory problems was hearing sensitivity, where certain loud noises, such as a school bell, hurt my ears. It sounded like a dentist drill going through my ear.”
- Temple Grandin
“One of my sensory problems was hearing sensitivity, where certain loud noises, such as a school bell, hurt my ears. It sounded like a dentist drill going through my ears.”

- Temple Grandin

**Acoustic shock**

- Constellation of symptoms that develop following exposure to sudden, unexpected sound:
  - Pain in ear, neck
  - Hyperacusis, hypervigilance
  - Tinnitus
  - Ear fullness/blocked sensation
  - Sound distortion
  - Vestibular disturbances
  - Numbness, burning, tingling
  - Headaches
  - SNHL occ. (not nec. 4 kHz notch)

- Mild to severe
- Temporary to permanent

**Interpreting services and ASI**

Interpreters surveyed (N=1035) reported:

- 67% had experienced an acoustic event (loud, unexpected, short duration)
- 47% had experienced symptoms of ASI

Mild to severe
Temporary to permanent

Similar presentation of symptoms to those of call centre workers

77% of incidents not officially reported (“Part of the job”)

Interpreters often not sure where to report, who to see about acoustic shock symptoms

May not be recognised by employers, etc.

Fear that acknowledging acoustic shock will hinder job prospects

COVID-19: loss of quality of sound due to poor connectivity, movement between both interpreter and person speaking
ASSESSMENT

When to assess

Estimated prevalence is 9-10% of the general population
Are we asking the right questions in our case histories?
Do people with DST avoid audiology clinics?
Shame or embarrassment?
Only seek help when in crisis?

Behaviours & complaints

Overuse of hearing protection
Avoidance behaviours
Social isolation
Loud sounds increase tinnitus
Pain or discomfort from sound
Amplification —>
Headache, discomfort
Increased tinnitus
"Can't wait to get them out."
Concerns

- Fackrell et al. (in progress) surveyed over 300 PWH –
- Fear (of damage or the unknown, fear for the future)
- Reduced quality of life
- Pain
- Restricts activities
- Avoidance

Interview components

- History
- Onset
- Comorbidities
  - Hearing loss
  - Tinnitus
  - Mood
  - Trauma
  - Head/neck injury/dysfunction
- What sounds are problematic?
- In what context?
- What reaction do they provoke?
- “How much of a problem is this for you?”

<table>
<thead>
<tr>
<th>Sound</th>
<th>Context</th>
<th>Who</th>
<th>Reaction</th>
<th>Problem (1-10)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dishes clattering</td>
<td>Kitchen at home</td>
<td>Anyone</td>
<td>Painful</td>
<td>5</td>
</tr>
<tr>
<td>Wind</td>
<td>Riding bike</td>
<td>Me</td>
<td>Tinnitus increases</td>
<td>3</td>
</tr>
<tr>
<td>Children screaming</td>
<td>Baby sitting</td>
<td>Grandchildren</td>
<td>Discomfort</td>
<td>8</td>
</tr>
<tr>
<td>Chewing</td>
<td>Mealtime</td>
<td>My partner</td>
<td>Furious</td>
<td>10</td>
</tr>
</tbody>
</table>
Measurement tools

- Questionnaires
  - No single questionnaire captures the diversity of experiences of PWH
  - Not sensitive to treatment effects
- Adults
  - Hyperacusis Questionnaire (Wraith, 2002)
  - Multiple-Activity Scale for Hyperacusis or MASH (Dauman & Bouscau-Faure, 2001)
  - Sounds Sensitive Tinnitus Index or SSTI (Greenberg et al., 2016)
  - Sound Tolerance Interview and Questionnaire Instrument (Sherlock & Formby, 2017)
- Children
  - One in development (University of Nottingham)

Loudness discomfort levels

<table>
<thead>
<tr>
<th>Pros</th>
<th>Cons</th>
</tr>
</thead>
<tbody>
<tr>
<td>Not uncomfortable if done properly</td>
<td>Risks causing pain</td>
</tr>
<tr>
<td>Allows monitoring of treatment</td>
<td>Might lose the patient’s trust</td>
</tr>
<tr>
<td>Allows feedback to patient</td>
<td>Results show a lot of variability</td>
</tr>
<tr>
<td>Facilitates demographic studies</td>
<td>Might trigger tinnitus</td>
</tr>
<tr>
<td>Know when to avoid acoustic reflex measurement</td>
<td>No global standard: &lt;90dB, &lt;100dB, dynamic range &lt;55-60dB</td>
</tr>
<tr>
<td>Variable between and within individuals</td>
<td>(Stephens et al., 1977)</td>
</tr>
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</table>

Effects of SNHL on LDLs

- Kamm et al., 1978
LDL procedure
Consistent instructions
Stop just before it gets uncomfortable
Test at all test frequencies
Duplicate runs (better, worse)
May need to proceed in 1-2 dB steps

What is important to the patient?
“What do you hope to get out of your appointment today?”
Motivational counselling tools
Road map
There are no clear clinical guidelines...

Patients will have developed their own coping strategies

- Avoidance
- Asking others to stop making sounds
- Hearing protection
- Music, headphones
- Coping strategies may be maladaptive

They may or may not be happy with their strategies

- “My world has gotten very small.”
- “We eat soup every night.”
- “I have my own company and have created a ‘misophonia friendly’ office space.”
- “I hide everything that people might fidget with when friends come over.”
- “I’ve bought quiet utensils and dishes for my friends that have me over.”
- “I’m happy but my life is very restricted.”
What treatments are PWH offered?

- Tinnitus retraining therapy
- Sound therapy
- Cognitive behavioural therapy
- Education and reassurance
- Relaxation strategies
- Mindfulness

Counselling

- Validation
- Explanation of mechanism
- “Safe” sounds
- Review of proper use of hearing protection
- Gradual and controlled sound exposure
- Gradual reduction of inappropriate ear plug use

Weaning off hearing protection

- Sound level meter - opportunity for obsessive checking of dB
- Instead
  - “Arm’s length” rule
  - How are others reacting to the sound?
  - Safe environment first
  - Can have HPD on-hand in case
  - Non-linear passive (Dancer & Hamery, 1998) or active electronic ear plugs (Etymotic)
  - Generic musician ear plugs
Sound desensitisation

- **Desensitisation**
  - Listening to sound just below the level that causes discomfort
  - Gradually increasing the level

- **Exposure**
  - Identify bothersome sounds
  - Gradually increase exposure duration, intensity (e.g. moving closer)
  - Careful not to over-expose
    - OST, tinnitus may temporarily increase

Sound therapy

- **Recalibration**
  - Using a constant level of white noise to reset the central auditory gain
  - Reduces contrast between background and bothersome sounds

- **Ear-level instrument**
  - Sound generator
  - Tinnitus combination instrument

- **Restricted use of amplification initially**
  - Safe environments
  - Gradually increase to target gain

Other options:

- White noise through ear buds on shoulders
- Sound machine at night
Rationale for sound therapy

Possibly resets auditory gain
“Sound shield”
Allows user to experience sound environments without annoyance/pain/fear
Sense of control
Facilitates decreased use of hearing protection
Limited evidence

Hearing aid selection

Flexible output and compression settings
Improving acceptable noise level (ANL)
Noise reduction - can be reduced as sound tolerance (ST) improves
Direction and remote microphones
May need to begin with custom earmolds, move to domes
Multiple programs - incl. “safe” program for loud environment
Datalogging
Volume control

- Searchfield & Sevkaratnam, 2018

Combination instruments

Internally generated sound generator (SG) or Bluetooth streaming of SG
app also an option
SG can be used alone or in conjunction with amplification, phased out as ST improves
Let patient know what to expect wrt hearing function
Hypervigilance
• Increased startle response to sound
• Increased awareness of sound environments
• Sound therapy may impede user’s ability to monitor environment for potential “threats”

Formby et al. 2013
Modified TRT approach
4 groups of 9 subjects with hyperacusis
- SG with (1) and without (2) counselling
- Placebo SG with (3) and without (4) counselling
80% of full treatment group had 10 dB improvement in LDLs after 6 months
Subjects were better able to tolerate amplification
Evidence-informed practice

Scoping reviews

Adults
Fackrell et al. 2017

Children
Potgieter et al. 2020

Evidence:
Sound therapy

Components: Acoustic training sessions, TinniTool, cochlear implants, hearing aids

Adults
11 studies showed improved LDLs, HQ scores, improved ability to manage life

Children
Case study, WNGs: ability to cope much better

Evidence:
Tinnitus retraining therapy

Hyperacusis TRT: Educational counselling, guidance on avoidance behaviour, sound desensitization and sound enrichment

Adults
15 studies suggested SGs and counselling led to greater improvements
1 study showed no change in sleep, relaxation, concentration, work or social activities

Children
1 study showed significant improvement for 75% of participants after 2-3 months for most patients, after 6 months for remaining 25%
Cognitive behavioural therapy

Evidence-based thinking

Evidence: Cognitive behavioural therapy

Includes education, sound desensitization, relaxation, cognitive restructuring

Adults
One RCT showed significant differences in HQ and LDLs compared to baseline and wait list controls

Children
Case study – “Improvement seen 2 weeks later in resisting troublesome sounds”

Psychology

- Various psychological approaches have been suggested
- CBT most widely adopted
- Gentle stepwise approach recommended
- Finding a psychologist or registered counsellor with appropriate skill set may present a barrier
Misophonia treatment options

Counselling
- Validation
- Explanation of mechanisms
- Hope

Ear-level sound generators, headphones + MP3 to minimize perception vs. ear plugs
Desensitisation (controlled exposure) while engaged in pleasant activity
Psychological care: contextual contributors, CBT, MBSR
Peer support

Evidence for treatment

Low-level evidence for CBT and sound-based therapy (TRT)
Most CBT studies are case studies or small groups
Combined approach may be prescribed, evidence lacking

Feedback from PWM varies

“Sound generators were a game changer.”
“My auditory system is becoming less sensitive to sounds [as my hearing instruments are adjusted].”
“My hearing instruments help me to block out trigger sounds and cope with everyday environments.”
“[Cognitive therapy] was a nightmare for me... but I have heard of it working for other people.”
“It took time to find a physician that was non-judgemental and didn’t seem to blame me for having misophonia.”
Opportunities for improvement

- Increased training for HCP
- Standardization of Terminology
- Assessment tools
- Treatment protocols

COVID-19 and DST

Pros
- Easier to avoid difficult environments
- Virtual interactions – can control volume
- Telehealth – easier than in-clinic visits

Cons
- Noise from neighbours day and night
- Family members, esp. children at home

Accessibility

Accessibility is about creating communities, workplaces, educational institutions, and services that enable everyone to participate fully in society without barriers.
Accessibility
Most people with DST express a desire to re-engage with sound
Removing systemic barriers to engaging with sound for the aurally diverse
How can audiologists help?
Validation of patient experience
  Build awareness
    • Employers
    • Healthcare providers (OT, VocReh, ENT, GP…)
Advocacy

DST at school
Flexibility, control
Accommodations for misophonia
  No eating in the classroom
  Lunch spot?
  Exams in separate room
  Triggers:
  - Dishes
  - Vacuum
  - Family members
  - Neighbours
  - Construction outside

DST at home
Dishes
Vacuum
Family members
Neighbours
Construction outside
Modification of environment

Hyperacusis recognized as trigger for behaviours in ASD
Reduce and contain noise
Sound absorbing materials

<table>
<thead>
<tr>
<th>Booklets:</th>
<th><a href="http://www.kingwood.org.uk">www.kingwood.org.uk</a></th>
</tr>
</thead>
<tbody>
<tr>
<td>Green Spaces:</td>
<td>Outdoor Environments for Adults with Autism</td>
</tr>
<tr>
<td>Living in the Community:</td>
<td>Housing Design for Adults with Autism</td>
</tr>
</tbody>
</table>

DST at work

Occupational health and safety aims to prevent NIHL, maintain safety
85 dB Leq
Sound levels deemed "safe" may be intolerable to those with DST

Leq: 75.6 dBA
Lpeak (max): 118.4 dB

Work No. 569, Piano, 2006
Martin Creed
Hearing protection

Overuse of hearing protection common
Occupational hazard
Overuse may bring on hyperacusis (Gold et al., 2003)
Worker may use to feel safe in anticipation of noise

Electronic hearing protection
Provide sense of control
Demonstrate, provide letter

Electronic hearing protection
Provides mild amplification of ambient sounds < 82 dB A
Attenuates sounds above 82 dB A
Reduces tinnitus aware

Safety rating
Class A (Peltor Tactical)
Class B (Bilsom Impact)

Communication needs and HPD

Does the worker need to communicate via radio?

DBM to electronic HPD to improve SNR
Caveat — this places audio input directly at the ear
Acoustic shock risk? 
DST in the office
Flexibility, control
Separate work space
Calm environment
Lighting
Sound absorption
Limited clutter
“What do you need to be able to do your work?”

DST and healthcare
Audiological testing including ABR, acoustic reflexes, VEMPs
Ear suctioning, irrigation (manual removal preferred)
MRI - talk to your MRI technologists

DST at play
Entertainment – expectation that audience has a particular capacity for sound
Live music
Movies
Sound installations
Expectations
Experimental music
Bird and Person Dyning, 1975
Alvin Lucier

The world is noisy!
How do we help our patients to adapt if the world cannot accommodate?
- Warnings prior to entry
- Access to appropriate HPD
- Control
- Exit plan

Sound can be used as a tool
Positively
- To augment an emotional experience (movies, art installations, etc.)
- To create atmosphere in public and private spaces
Certain experiences and spaces may not be easily accessible to those with DST
Maliciously
- As a deterrent (“Mosquito”)
- Bullying → assault
- Crowd control
- Potential for negative and long-term consequences
Online resources
Reliability and quality of information on hyperacusis (rated out of 5)

<table>
<thead>
<tr>
<th>Resource</th>
<th>Rating</th>
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<tbody>
<tr>
<td>Action on Hearing Loss</td>
<td>4</td>
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<tr>
<td>ASHA</td>
<td>3</td>
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<tr>
<td>British Tinnitus Association</td>
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<td>Hyperacusis Focus</td>
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<td>Hyperacusis.net</td>
<td>2.5</td>
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<td>WebMD</td>
<td>2.5</td>
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<td>Wikipedia</td>
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Smith et al. (2020) AJA

Research questions

What is the most effective treatment approaches for
- children?
- Persons with autism
Which treatment approaches are best for various sub-types of hyperacusis or degrees of severity?
Which psychological therapy is most effective for hyperacusis? (CBT, counselling, mindfulness)
Which self-help interventions are effective?


Summary

Loudness tolerance problems are common if patients are asked the appropriate questions
Terminology is a work in progress
Aetiology still obscure in most cases
Strong link with tinnitus
Auditory system remains plastic throughout life
Sound based and psychological treatments
Aural diversity includes persons with DST
Audiologists can...

- Provide tools and counselling to manage and treat DST
- Help address systemic barriers encountered by persons with DST
- Promote prevention of acoustic environments that can produce or exacerbate DST

WATCH THIS SPACE...