Assessment and management of Acoustic Shock, Tonic Tensor Tympani Syndrome (TTTS), hyperacusis and misophonia

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Myriam Westcott
Definitions

- **Hyperacusis** =
  - an abnormal sensitivity or intolerance of certain sounds, which other people can tolerate
  - + a heightened sense of volume
  - + physical discomfort

- Sounds that are typically difficult to tolerate: loud/impact sounds, particularly if unexpected and in close proximity.

- Exposure to intolerable sounds causes panic, distress, anxiety, aural symptoms of pain, blockage etc.
Definitions

**Misophonia** =
a strongly aversive response to certain specific sounds, often made by other people, irrespective of their volume.

- It may develop when those sounds become perceived as an intolerable intrusion into one’s sense of personal space.

- Online forums - raised awareness, widespread.

- Misophonia not listed in the DSM-IV TR or ICD-10 systems, considered a form of OCD, currently being investigated re classification as a discrete psychiatric disorder.

- Exposure to intolerable sounds causes resentment, annoyance, anger, disgust, rage.
How do H and M develop?

H, M = result of a sense of threat from specific, everyday sounds.

Sounds evaluated in the subconscious as threatening are:
- judged as important
- highlighted
- appear to be boosted in volume
- transmitted to the more conscious parts of the brain
- become increasingly prominent

H and M can occur concurrently - but the nature of the threat from these sounds is different, so they are separate conditions.

H, M:
- can range from mild to severe to extreme
- has the potential to escalate, so that an increasing range of sounds become intolerable.
H:
Can develop in the belief that:
- ears/hearing are being damaged by intolerable sound exposure
- pre-existing aural symptoms will be aggravated: tinnitus, hearing loss, TTTS symptoms
- fear of: discomfort/pain; distress from intolerable sounds

M:
- disgust, rage etc
- fear/resentment of: experiencing these emotions; inability to control them following exposure

H, M - major impact:
- severe lifestyle restrictions
- auditory hypervigilance: constant environmental monitoring
- difficulty in managing high levels of emotions pre and post exposure
- difficulty in explaining this abnormal reaction to sound to others, including health professionals
H: Co-morbid conditions

H can develop with:

- Tinnitus, particularly if anxiety levels are high
- Acoustic shock
- Auditory pathway pathology eg: Meniere’s Disease, otosclerosis, Bell’s Palsy, perilymph fistula, superior canal dehiscence
- Autistic spectrum disorders
- Adverse reaction to some medications
- Psychiatric disorders
- Neurological injuries/disorders:
  - head injury, migraine
  - myalgic encephalomyelitis (chronic fatigue syndrome)
  - fibromyalgia, Lyme Disease
How do H, M develop in children?

Children:
- less control over their environment
- encouraged to be sociable, avoidance behaviours often discouraged
- kindergartens, schools are noisy - sounds are unpredictable
- anxious children are vulnerable to H development

M:
often stems from an aversive reaction to specific sounds made by family members in childhood or teenage years.

H and M:
Autistic spectrum disorders:
- readily overwhelmed by auditory information
- trying to identify and make sense of sounds
- difficulties in filtering out unimportant sounds
Secondary symptoms of H

- anger, anxiety, depression
- auditory hypervigilance
- reduced startle reflex
- Tonic tensor tympani syndrome (TTTS)
- enhanced awareness of tinnitus

If H has developed from or become exacerbated by a sound perceived as traumatic, TTTS symptoms consistent with acoustic shock disorder (ASD) may have developed.
Acoustic shock (AS) =
an involuntary response to an (emotionally) traumatic sound, leading to a characteristic cluster of neurophysiological symptoms in and around the ear(s).

The acoustic incident triggering AS is usually a loud/sudden/unexpected sound heard close to the ear(s).

AS symptoms are highly consistent, usually temporary.

For some, they can be:
- severe
- escalate
- persistent
- become permanently disabling (ASD)
Initial symptoms of acoustic shock

- Severe startle reaction with a head/neck jerk
- Stabbing pain in the ear
- Dull ache in the ear, can radiate to cheek, neck, arm
- Tinnitus, hyperacusis
- Sensation of fullness/blockage in the ear
- Sensations of burning, numbness, tingling in and around the ear
- Mild vertigo, nausea
- Muffled, distorted hearing (usually subjective with no measurable change in hearing)
- Shock response: shaking, crying, disorientation, headache, fatigue
ASD: Symptom development

If these symptoms persist, secondary symptoms may develop:

- **hyperacusis:** persistent symptoms exacerbated by loud/intolerable sound exposure

- **psychological symptoms:**
  - auditory hypervigilance
  - reduced startle reflex
  - anxiety, depression, adjustment disorder
  - post traumatic stress disorder/critical incident stress

- **trigeminal nerve irritability,** potentially leading to chronic trigeminal neuralgia, TMD
Call centre staff - vulnerable to AS, AS research has focused on this cohort.

BUT…..
Acoustic incidents can occur anywhere

AND
Many patients with tinnitus and hyperacusis also report some/many ASD symptoms.

Patuzzi, Milhinch and Doyle (2000) and Patuzzi (2002) considered:

- The primary cause of AS due to excessive middle ear muscle contractions from exposure to the acoustic incident.

Tensor Tympani Muscle
The tensor tympani reflex:

- variable threshold to sound, which can be reprogrammed downwards

- can be triggered without sound

- activated as part of the startle response, which becomes exaggerated with high stress/anxiety levels

- established protective function: contracts immediately before self-vocalisation

- considered to be primarily responsible for aural symptoms secondary to TMJ dysfunction/TMD.
Tonic tensor tympani syndrome (TTTS)

Ramirez et al, 2008:

- TTTS triggers physiological reactions from tympanic membrane tension and alterations in middle ear ventilation (tympanic flutter, sensation of blockage/fullness, “muffled” hearing).

- TTTS can lead to irritability of the trigeminal nerve, as well as other cranial and cervical sensory nerves in and around the ear (neuralgic pain, burning, numbness, tingling and tenderness in and around the ear).

The cervical system is a sensory organ. Muscular tension and pain in the neck can result in:

- dizziness (unsteadiness, disequilibrium)
- nausea
- aural fullness
- TMJ pain.

Central pain sensitisation can develop from chronic pain, leading to an expansion of the perceived peripheral pain (muscle trigger point development in neck, shoulder and arm).
Neural plasticity, central pain sensitisation

Herta Flor
(TRI conference, Stresa 2009):

- Duration of pain, rather than intensity, increases risk of central sensitisation.

- Chronic pain: descending inhibition reduces.

- Anxiety, depression, catastrophising evokes enhanced pain perception.

- Unpredictable pain lowers pain thresholds. People with no control over pain unable to activate an inhibitory descending circuit.

If unpredictable, everyday sounds hurt, and a person with H can’t avoid these sounds........
Multi-clinic TTTS symptom prevalence study: participants

- **Tanit Ganz Sanchez** (43 patients), Sao Paolo, Brazil
- **Clarice Saba** (72 patients), Salvador, Brazil
- **Isabel Diges** (53 patients), Madrid, Spain
- **Mary O’Keefe** (10 patients), Auckland, New Zealand
- **Alison Chiam** (35 patients), Vincentia, Australia
- **Celene McNeill** (15 patients), Sydney, Australia
- **Tricia Sharples** (12 patients), Hobart, Australia
- **Ross Dineen** (21 patients), Melbourne, Australia
- **Myriam Westcott** (92 patients), Melbourne, Australia
Multi-clinic TTTS symptom prevalence study: data collection

- Data collected on all consecutive T and H patients seen over the survey period

- 3 groups: T tinnitus only, T+H tinnitus + hyperacusis, H hyperacusis only

- degree of severity of T/H (mild/moderate/severe)

- whether the T, H had been triggered by an acoustic incident

- age, gender, hearing loss (4 frequency average)

- presence of symptoms consistent with TTTS in one or both ears

- whether these symptoms developed from, or were exacerbated by, exposure to loud/intolerable sounds
Multi-clinic TTTS symptom prevalence study: data collection

All patients with symptoms consistent with TTTS cleared of:

- outer/middle/inner ear pathology
- TMJ dysfunction or pathology
- if symptoms were unilateral, of retrocochlear pathology.

Patients whose only symptoms were:

- headache
- vertigo
- muffled/distorted hearing were not considered to have symptoms consistent with TTTS.

Muffled/distorted hearing was excluded as a symptom consistent with TTTS in patients with a hearing loss.
Multi-clinic TTTS symptom prevalence study: results

Total 345 patients (52% male, 48% female)

49.3% T group (170/345)
42.3% T+H group (146/345)
8.4% H group (29/345)

Hyperacusis present in:
- 50.7% (T+H, H groups) of total sample
- 46.2% of those with tinnitus (T+H group)

60.0% (207/345) of total sample had at least one symptom consistent with TTTS

- 40.0% (138/345) no symptoms
- 12.5% (43/345) one symptom
- 47.5% (164/345) $\geq$2 symptoms
## Most common symptoms

<table>
<thead>
<tr>
<th>TTTS Symptom</th>
<th>T Group n=170</th>
<th>T+H/H Group n=175</th>
</tr>
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<tbody>
<tr>
<td>Aural fullness</td>
<td>13% (22/170)</td>
<td>53% (92/175)</td>
</tr>
<tr>
<td>Headache</td>
<td>14% (23/170)</td>
<td>29% (50/175)</td>
</tr>
<tr>
<td>Disordered balance</td>
<td>12% (20/170)</td>
<td>29% (50/175)</td>
</tr>
<tr>
<td>Dull ache in ear</td>
<td>9% (16/170)</td>
<td>29% (50/175)</td>
</tr>
<tr>
<td>Tympanic flutter</td>
<td>10% (17/170)</td>
<td>28% (49/175)</td>
</tr>
<tr>
<td>Sharp pain in ear</td>
<td>8% (13/170)</td>
<td>27% (48/175)</td>
</tr>
<tr>
<td>TMJ pain</td>
<td>15% (25/170)</td>
<td>25% (44/175)</td>
</tr>
<tr>
<td>Pain – side of neck</td>
<td>15% (25/170)</td>
<td>25% (44/175)</td>
</tr>
<tr>
<td>Muffled hearing</td>
<td>8% (14/170)</td>
<td>17% (29/175)</td>
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</tbody>
</table>
Results

- Statistical evidence that symptoms are clustered ie: belong to a syndrome

- Symptoms consistent with TTTS common in T patients ($\geq 1$ symptoms in 40.6%), particularly if they have developed H ($\geq 1$ symptoms in 81.1%)

- The more severe the T/H, the more likely the patient will have $\geq 2$ symptoms consistent with TTTS

- Symptoms can be aggravated by intolerable sound exposure, particularly for those with H

- Unilateral preponderance of symptoms in patients with:
  - unilateral H
  - AS triggered by unilateral acoustic incident exposure

- Results consistent with an alternative aetiologic pathway triggering TTTS as a primary phenomenon in T, H and ASD patients
  - distinct from secondary TTTS in patients with TMD and/or TMJ dysfunction.
Results: AS (acoustic incident trigger + TTTS symptoms)

- 24.1% of total sample identified an acoustic incident trigger for their T/H onset

- Of those, 81.9% reported ≥1 symptoms consistent with TTTS, compared to 53.1% of patients without an acoustic incident trigger (p<0.001)

- 19.7% of total patients had AS

- AS reported in all countries: Aus/NZ 26.3%; Spain 13.2%; Brazil 12.4%

- 83.8% of AS patients had H, 41.2% of non-AS group had H (p<0.001)

- 97% of patients with a unilateral acoustic incident trigger for their AS had ≥1 symptoms consistent with TTTS to a greater degree on the affected side (p<0.001)
Preliminary results presented at Xth International Tinnitus Seminar, Brazil March 2011,
- selected by the Scientific Committee to receive the inaugural Jack Vernon Award for originality, innovation, study design, analysis and ultimate impact on the field of tinnitus.

This study presented at the 5th International TRI Conference on Tinnitus, Buffalo, USA, August 2011
TTTS: clinical observations

Explain why:
- H patients report pain after exposure to sounds they find intolerable, but others tolerate well.
- Severe H patients are in frequent/constant pain, exacerbated by intolerable sound exposure.

TTTS symptoms can appear in patients with an (often involuntary) “need to protect” their ears.

H desensitisation strategies reduce TTTS symptoms.

Understanding and explaining TTTS provides reassurance, gives a strong framework for desensitisation.

Tracking TTTS symptoms is a non-invasive clinical indication of the efficacy of a H desensitisation program.
Evaluation: H, M, ASD, TTTS

Detailed history:
- I analyse in depth with each patient how sounds changed from being tolerable to intolerable, and pattern of escalation.
- Triggers, stress levels at time of onset?
- ASD: acoustic incident details, immediate/persistent symptoms, workplace management, prior acoustic incident exposure, malingering
- Which sounds are intolerable? Symptoms (TTTS, emotional) following intolerable sound exposure?
- Prior otological and psychological history, screening for anxiety/depression.
- Clear diagnosis: H, M, ASD, TTTS

Questionnaires:
- H: Khalfa, HRQ
- M: The Amsterdam Misophonia Scale (A-MISO-S)
ASD or TMJ (jaw joint) disorder?

ASD symptoms often misdiagnosed as due to TMJ dysfunction causing TMD.

**Differential diagnosis:**
- careful history taking: identification of an acoustic incident at the time of symptom onset
- presence of H, symptom aggravation following intolerable sound exposure
- symptom development/escalation over time.

With ASD, any TMD symptoms are secondary to aural symptoms. With TMD from TMJ dysfunction, aural symptoms are secondary to TMJ symptoms.
Audiological assessment: H, ASD

- PTA *done with extreme care:* ascending technique
- No LDLs, minimise suprathreshold testing
- Tympanometry (*without stapedial reflex measurement*)

ASD:
- Muffled/distorted hearing common, usually subjective
- If hearing loss present: mid and/or low frequency sensorineural, temporary or permanent
- Normal tympanometry, normal clinical exam of ear, normal MRI
Audiological evaluation and management of ASD

- Comprehensive audiological assessment
- Rapid diagnosis based on detailed history and symptom cluster
- Providing acknowledgement and reassurance; an understanding of ASD; counselling
- Overseeing changes to work duties
- Referral to an ENT specialist, psychologist, pain management clinic.
- Detailed report including ASD diagnosis: may need to defend in court, helpful as validation for patient, guides other health professionals
- Long term rehabilitation strategies for tinnitus, hyperacusis and persistent TTTS symptoms.
Onward Referrals

**ENT Specialist:**
- Vertigo/balance disturbances – to exclude fistula
- Severe TTTS symptoms – to exclude aural ear pathology
- To support diagnosis of ASD (necessary for 3rd party funding)

**Psychologist/psychiatrist:**
- anxiety, depression and trauma treatment. Severe ASD/hyperacusis can lead to PTSD, critical incident stress, adjustment disorder
- PTSD patients can have ASD/hyperacusis

**Pain Specialist:**
ASD/H patients with constant pain
H, TTTS, M desensitisation – how?

- I encourage patients not to blame themselves (or others) for developing H, M, TTTS, ASD. M often stems from an aversive reaction to certain sounds made by family members.

- H,M patients need to acknowledge their sound intolerance as an abnormal reaction they want to change – people can get stuck at this point if they justify it as reasonable.
H, M, TTTS Desensitisation Therapy

- TRT based: An explanation of the peripheral and central auditory pathway, including hearing test results and the neurophysiological basis of H, M, TTTS, and tinnitus awareness/distress.

- Cognitive behavioural therapy (CBT):
  - reframing destructive thoughts/beliefs
  - auditory hypervigilance management

- Sound enrichment strategies
Neurophysiological model of tinnitus distress and hyperacusis (P Jastreboff)
Cognitive behavioural therapy

I teach my patients to recognise, challenge, reframe any irrational beliefs that may contribute to and exacerbate their H, M, TTTS.

Many irrational beliefs seem logical, and changing them can seem counter-intuitive:

- “Intolerable sounds hurt/increase my tinnitus, so they must be damaging my ears.”

- “My ears are more sensitive than others, because I’ve got tinnitus/noise damage/ear pathology already. I need to protect them to avoid further damage.”

- “I can’t lead a normal life because I have to avoid noisy places.”

- “I have super hearing, I can hear sounds others can’t.”
Auditory hypervigilance management

- Accept that intolerable sounds will inevitably be present at some time

- Focusing on intolerable sounds inhibits desensitisation. Actively listening for them brings them into conscious awareness, perpetuates threat.

- As a result, the brain will continue to highlight these sounds: they appear louder, can’t be masked by tolerable sounds, enhances potential for escalation.

- Identify or create a “safe space”

- Distraction strategies

- Mindfulness strategies
Sound enrichment

TRT protocol.

Additionally, I counsel my patients to use sound:

- as a barrier or shield to intolerable sounds
- to promote a sense of detachment from threatening sounds
- to help create and define a portable “safe space”
Protecting the ears – yes or no?

Yes, but (only) to maintain or expand lifestyle horizons.

**Ear plugs**: foam, solid silicon, filtered musician’s earplugs.

**Hearing aids**: carefully programmed
- amplify softer sounds to: compensate for occlusion; reduce tinnitus awareness; support a hearing loss, if present.
- higher compression to reduce startle to impact sounds, particularly for high frequencies.
- low MPO to act as a filter to loud sounds.
- opportunity for desensitisation by gradually increasing MPO and reducing compression.
Management of emotions

I encourage my patients to:

- identify emotions felt after exposure to intolerable sounds: panic, fear, anxiety, distress, anger, intrusion, invasion of “my” space.

- accept they are having these emotions, acknowledge they want to cope better with them.

- not blame themselves for having them or deny they are occurring. They are aiming not to suppress the emotions but to understand and manage them more effectively.

- Stress/panic/anger management: breathing techniques, imagery, active relaxation, mindfulness meditation.

- Grief counselling: Lifestyle limitations, loss of confidence and ability to mix in groups, the effect of H, M on relationships, career/employment limitations – loss of financial security, coping in the future.
Case study: DB

- Call centre operator for 6+ years. Used monaural headset on right ear.

- First acoustic incident exposure via headset several years prior.

- Second acoustic incident. Immediate symptoms: severe fright reaction, cognitive disorientation.

- Resumed headset duties after a few minutes.

- Several days later, noticed intermittent pain in right ear.
Steady escalation in right aural pain: spasmodic, stabbing pain ‘like jabbing a needle in my ear’

Other symptoms developed:
- intermittent right aural fullness
- bilateral tinnitus, greater in right ear
- frequent nausea
- H to loud voices, sudden unexpected sounds
- burning sensation in and around right ear
- sensitivity to touch in and around her right ear, although able to use ear plugs.
Consulted 3 ENT specialists: CT scan, MRI clear of pathology


ASD diagnosed.

Therapy:
- Detailed explanation of the peripheral and central auditory pathway, including neurophysiological basis of ASD, TTTS, H and tinnitus distress
- CBT distraction strategies to reduce tinnitus awareness/auditory hypervigilance
- Sound enrichment strategies to support H/TTTS desensitisation and tinnitus habituation
- Sleep and stress management
H reduced. DB determined to keep working and not to restrict lifestyle. Lack of workplace support added to stress.

- Fitted with solid and ER25 ear plugs.

- All symptoms persisted but pain the most distressing symptom:
  - constant dull ache in and around ear
  - frequent burning sensation in and around right ear
  - frequent stabbing pain in ear.

- Referred to pain management clinic.
Pain management clinic evaluation

- Diagnosis: myofascial pain syndrome as a result of ASD.

- Pain enhanced by development of muscular trigger points around right ear extending to cervical/shoulder girdle and bilateral upper limb muscles. Managed by physiotherapy exercises/self massage.

- Spread of trigger points consistent with central nervous system pain pathway sensitisation ("a scientifically proven, organic change in her pain system effectively lowering her pain threshold").

- Endep prescribed- ? mild benefit noticed. Medication changed to Lyrica (Pregabalin anticonvulsant, used to treat pain caused by nerve damage), has been successful in reducing DB’s pain levels.
4 years later

- 14 months after injury, offered alternative job, much less stressed. Now retrenched, trying other employment.

- H: coping with moderate everyday sounds; impact/complex/loud noises remain difficult to tolerate

- Tinnitus gone, DB attributes this to successful use of therapeutic sound. Stress reduction also likely to be a factor.

- Aural fullness gone. Aural pain persisted, although at a reduced level (down to 30% most of the time).

- Symptoms fluctuate, depending on stress levels and H. If severe, sensation of tympanic fluttering noticed.

- Stopped taking Lyrica – led to a major increase in pain levels.

- Family crisis: inadvertent use of a mobile phone on right ear, leading to a persistent increase in TTTS symptoms, pain levels increased and escalated to radiate down her right arm.

- DB continues to use TRT/CBT strategies to manage symptoms.
Conclusion

I consider that ASD, H, M, TTTS need to be:

- acknowledged as legitimate conditions
- diagnosed and demystified to patients

The potential severity and persistence of ASD symptoms has significant clinical, occupational and medico-legal implications:

- With the global growth of call centres, tinnitus clinicians are increasingly likely to encounter ASD.

- Acoustic incidents can be expected to occur in environments such as war zones….increasing incidence of tinnitus + PTSD in American war veterans.