

Hyperacusis



Hyperacusis

- A condition marked by super sensitive hearing.
- Often environmental sounds are so loud to them it is painful.

Edelson, S. (1994 Spring). Your Questions Answered. Perspectives of the Orton Dyslexia Society, 20 (2), p. 7.



Hyperacusis



“Hyperacusis has been defined as ‘unusual tolerance to ordinary environmental sounds’ and more pejoratively, as ‘consistently exaggerated or inappropriate responses to sounds that are neither threatening nor uncomfortably loud to a typical person.’ Common to both is the implication that the experience can be evoked by sounds of low intensity and that sounds in general, rather than specific sounds are problematic” (p. 582)

Baguley, D.M. (2003). Hyperacusis. Journal of the Royal Society of Medicine, 96 (12), 582-585.

Vestibular Hyperacusis



“In *vestibular hyperacusis*, exposure to sound can result in falling or loss of balance or postural control...Some of the same reactions as with cochlear hyperacusis can also occur, along with sudden vertigo and nausea” (p. 1 of 2).

- Other symptoms: loss of consciousness, confusion, fatigue, headaches

Author (May 1, 2010). Vestibular Hyperacusis. From Vestibular Association (VEDA)
Website: www.vestibular.org/vestibular-disorder/specific-disorders/vestibular-hyperacusis, p. 1 to 2.

Vestibular Hyperacusis



- Causes:
 - Head/brain injury, chemical exposure, heart/artery disease, autoimmune disorders
- Treatment:
 - Low salt diet, anti-nausea drugs, anti-inflammatory drugs.

Author (May 1, 2010). Vestibular Hyperacusis. From Vestibular Association (VEDA) Website: www.vestibular.org/vestibular-disorder/specific-disorders/vestibular-hyperacusis, p. 1 to 2.

Loudness Recruitment



“Loudness recruitment describes an experience associated with cochlear hearing loss and specifically with dysfunction of the outer hair cells of the organ Corti: with a rising sound level, the perceived loudness increases faster than normal”. (p. 582)

Baguley, D.M. (2003). Hyperacusis. Journal of the Royal Society of Medicine, 96 (12), 582-585.

Loudness Recruitment



“This phenomenon occurs because at some decibel level, the normal hair cells adjacent to the damaged hair cells (corresponding to the frequency of a hearing loss) are ‘recruited’. At the decibel level at which these normal hair cells ‘kick in’, perceived loudness shoots up rapidly, causing discomfort”. (p. 2 of 3)

Author (2010). Types of Sound Sensitivity. From Hyperacusis Network website:
www.hyperacusis.net/hyperacusis/4+types+of+sound+sensitivity/default.asp, p
1 to 3.

Loudness Recruitment



- Usually occurs typically occurs in people with high frequency hearing loss over the age 40.
- They have an abnormal perception of loudness and are overwhelmed by it.

Author (2010). Suppliment. From the Hyperacusis Network website:
www.hyperacusis.net/hyperacusis/suppliment/default.asp, p. 1 to 25.

Misophonia and Phonophobia

“Misophonia and Phonophobia can be defined as abnormally strong reactions of the autonomic and limbic systems resulting from enhanced connections between the auditory and limbic systems” (p. 582)

- Phonophobia = Fear of sounds due to an emotional pairing. Usually term used to describe lack of sound tolerance due to migraine headaches.
- Misophonia = Dislike of sounds due to an emotional pairing

Jastreboff, M.M., and Jastreboff, P.J. (June 18, 2001) Hyperacusis. From:
www.audiologyonline.com/articles/article_detail.asp?article_id=291, p. 2 of 3.

Baguley, D.M. (2003). Hyperacusis. Journal of the Royal Society of Medicine, 96 (12), 582-585.

Note!



- Most people will find sounds at 120 dB or higher painful.
- The maximum comfortable loudness a person can tolerate depends on the individual.
- It can also depend on your mood and the type of sound how well you can tolerate it.
 - The more stressed you are the less you can tolerate sound.
- Some sounds are intolerable to everyone (i.e. fingers scratching a chalkboard)

Baguley, D.M. (2007). Hyperacusis and Other Forms of Sound Tolerance. British Tinnitus Association. From website: www.tinnitus.org.uk.

Causes of Hyperacusis

- ❖ Note: In most cases no cause can be found.
- ❖ Facial nerve dysfunction
- ❖ Lyme disease
- ❖ William's Syndrome: 96% report hyperacusis
- ❖ Middle cerebral aneurysm
- ❖ Multiple sclerosis
- ❖ Migrainous cerebral infarction
- ❖ Ear surgery*



Baguley, D.M. (2003). Hyperacusis. Journal of the Royal Society of Medicine, 96 (12), 582-585.

*Baguley, D.M. (2007). Hyperacusis and Other Forms of Sound Tolerance. British Tinnitus Association. From website: www.tinnitus.org.uk.

Causes of Hyperacusis

- ❖ 5 hydroxytryptamine dysfunction: This helps modulate auditory gain and determining the importance of particular sounds.

Baguley, D.M. (2003). Hyperacusis. Journal of the Royal Society of Medicine, 96 (12), 582-585.

- ❖ Temporal Mandibular Joint Syndrome (TMJ)?

Author (2010). Suppliment. From the Hyperacusis Network website:
www.hyperacusis.net/hyperacusis/suppliment/default.asp, p. 1 to 25.

Causes of Hyperacusis

“Specifically, improper function of the cochlear hair cells may result in a hearing loss secondary to the failure of these cells to propagate proper signals through the auditory centers. In response to an incongruous neural message, higher auditory cortical centers may adopt and remodel transmitted sound. This neuroplasticity may lead to an increased perception of volume in the auditory cortex (hyperacusis) and the perception of phantom sounds (tinnitus)” (p. 472)

Nelson, J.J., and Chen, K. (July, 2004). The Relationship of Tinnitus, Hyperacusis, and Hearing Loss. Ear, Nose and Throat Journal, 83 (7), 472-476.

Hyperacusis



- Poll of 65 AD/HD adults:
 - 50% reported hypersensitive hearing

Johnson, M.J. (1998) Having ADD and Being Hypersensitive: Is There A Connection?.

From website:<http://www.add.org/articles/hypersen.html>

Asperger's Disorder and Hyperacusis



- 70 to 80% of those with Asperger's Disorder have some form of Hyperacusis.
- The three types of sounds people with Asperger's Disorder have difficulty with:
 - Sudden Unexpected Noises – dog bark, school fire alarm, etc.
 - High Pitched Continuous Noises - electric motors. toilets, etc.
 - Confusing Multiple Sounds – shopping mall, sporting event, etc.

Attwood, T. (2007). The Complete Guide to Asperger's Syndrome. Philadelphia, PA: Jessica Kingsley, p. 221.

Hyperacute Hearing



Some say that those with Autism Spectrum Disorders who are hypersensitive to sound have “hyperacute hearing”, instead of hyperacusis because they are born with it and only certain sound frequencies cause them problems.

Author (2010). Types of Sound Sensitivity. From Hyperacusis Network website:
www.hyperacusis.net/hyperacusis/4+types+of+sound+sensitivity/default.asp, p 1 to 3.

Williams Syndrome and Sensitive Hearing

- Williams Syndrome is also known as Williams-Beuren Syndrome
- “Williams syndrome is characterized by cardiac defects, varying degrees of physical and developmental delay, stellate eye pattern, possible elevated serum calcium level, and elfin/pixie facial features”(p. 339)
- 95% of the people with Williams Syndrome have hyperacusis and 61% had significant otitis media as infants and small children.

Klein, A.J, Armstrong, B.L., Greer, M.K., and Brown III, F.R. (May, 1990). Hyperacusis and Otitis Media in Individuals with Williams Syndrome. Journal of Speech and Hearing Disorders, 55, 339-344.

Williams-Beuren Syndrome (WBS)

“To begin with, individuals with WBS experience strong aversion to certain types of sounds, independent of their loudness. The sounds tend to be spectrally broad-band sounds, such as those emanating from motors, fans, fireworks, and thunder. Compared to typically developing children and children with DNS (sic. Downs Syndrome) or Autism, the WBS individuals are more than three times as likely to have suffered from auditory aversions, with 91% of respondents reporting this (compared to 27% of individuals with Autism, and fewer than 7% of individuals with DNS or normal controls)”. (p. 349)

Levitin, D.J., and Bellugi, U. (2006). Rhythm, Tembre and Hyperacusis in Williams,-Beuren Syndrome. In C. Morris, H. Lenhoff, and P. Wang (Eds.), Williams-Beuren Syndrome: Research and Clinical Perspectives. Baltimore, MD: John Hopkins Press, 343-358.

Williams-Beuren Syndrome (WBS)

“WBS people may suffer from four different auditory abnormalities: lowered uncomfortable loudness levels, hyperacusis, auditory facinations, and auditory aversions. We argue for some neural basis for some of these behaviour may be in hyperexcitability of cortical neurons...WBS tend to use different regions of their brains for processing music and noise, with particular emphasis on amygdala activation”. (p. 354)

Levitin, D.J., and Bellugi, U. (2006). Rhythm, Tembre and Hyperacusis in Williams,-Beuren Syndrome. In C. Morris, H. Lenhoff, and P. Wang (Eds.), Williams-Beuren Syndrome: Research and Clinical Perspectives. Baltimore, MD: John Hopkins Press, 343-358.

Williams-Beuren Syndrome (WBS)

“Hyperacusis in Williams syndrome (WS) is associated with a high-frequency hearing loss resembling the configuration of noise induced hearing loss. The hyperacusis and hearing loss in WS may stem from a deficiency in the acoustic reflex resulting from auditory nerve dysfunction. Additional mechanisms that may mediate hyperacusis in WS and should be evaluated in future studies include recruitment, malformation of the facial canal, and haploinsufficiency of the elastin gene”. (p. 390)

Gothelf, D., Farber, N., Apter, A., and Attias, J. (February, 2006). Neurology, 66 (3), 390-395.

Williams-Beuren Syndrome (WBS)



“Even as toddlers, children with William’s Syndrome are extraordinarily responsive to music...” (Oliver Sacks, 2007, p. 219)

Sacks, O. (2007). Musicophilia: Tales of Music and the Brain. New York, NY: Alfred A. Knopf, 19.

Tonic Tensor Tympani Syndrome (TTTS)

“In many people with hyperacusis, and increased activity develops in the tensor tympani muscle in the middle ear as part of the startle response to some sounds. The lowered reflex threshold for tensor tympani contraction is activated by the perception/anticipation of loud sound, and is called tonic tensor tympani syndrome (TTTS). In some people with hyperacusis, the tensor tympani muscle can contract just by thinking about loud sound. Following exposure to intolerable sounds, this contraction of the tensor tympani muscle tightens the ear drum, which can lead to symptoms of ear pain/a fluttering sensation/a sensation of fullness in the ear (in the absence of any middle or inner ear pathology). (Continued)

Tonic Tensor Tympani Syndrome (TTTS)

“...it does not harm the ear to experience TTTS, and even though the TTTS symptoms can seem as if the ear is being damaged by some sounds, this is not the case”. (p. 2 of 3)

- **Treatment of TTTS often involves sound desensitization and habituation wearing sound generators at low volume.**

Westcott, M. (2010). Tensor Tympani Syndrome. From the Hyperacusis Network website:
www.hyperacusis.net/hyperacusis/tensor+tympani+syndrome/default.asp. P1 & 2, of 3.

English Language Questionnaire for Hyperacusis

Khalifa, S., Dubal, S., Veillet, E., Perez-Seliaz, F., Jouvent, R, and Collet, L. (2002). Psychometric Normalization of a Hyperacusis Questionnaire. Otorhinolaryngology, 64, 438-442.



Sensory Sensitivity Symptoms in Asperger's Disorder

- Sensory Profile Checklist-Revised (SPCR): From - Bogdashina, O. (2003). Sensory Perceptual Issues in Autism and Asperger's Syndrome: Different sensory Experiences, Different Perceptual Worlds. London, England: Jessica Kingsley.
- Sensory Behaviour Schedule (SBS): From - Harrison, J., and Hare, D. (2004). Brief Report: Assessment of Sensory Abnormalities in People with Autism Spectrum Disorders. Journal of Autism and Developmental Disorders, 34, 707-730.

Prevalence of Hyperacusis

- Two Swedish studies indicated 8 to 9% of the population has hyperacusis.
- 40% of those with tinnitus have hyperacusis
- 86% of those with hyperacusis have tinnitus
- 48% of those with loudness recruitment complain of problems with attention as well as emotional and social concerns.

Baguley, D.M. (2003). Hyperacusis. Journal of the Royal Society of Medicine, 96 (12), 582-585.

Prevalence of Hyperacusis

- Most researchers believe the worldwide prevalence of hyperacusis is about 2%.

Baguley, D.M. (2007). Hyperacusis and Other Forms of Sound Tolerance. British Tinnitus Association. From website: www.tinnitus.org.uk.



Hyperacusis: Diagnosis



- Go to a “World Class” university medical center (i.e., Harvard Medical School, Vanderbilt Medical School, Mayo Clinic, UCLA Medical School, etc.) and have the following:
 - A thorough medical examination
 - A very detailed history taken (i.e., medical, family, educational, audiological, work, social, etc)
 - Thorough ear examination
 - Thorough hearing examination
 - Thorough hyperacusis examination that includes a loudness discomfort test.

Baguley, D.M. (2007). Hyperacusis and Other Forms of Sound Tolerance. British Tinnitus Association. From website: www.tinnitus.org.uk.

Good Book On Hyperacusis

Baguley, D., and Andersson, G. (2007).
Hyperacusis: Diagnosis, Mechanisms and
Therapies. London, England: Plural.



Treatment of Hyperacusis

- Some use exposure to “pink noise” with the hope that it will desensitize them to their hyperacusis.
- Pink Noise is similar to white noise.
- CDs with such sounds are available from the Hyperacusis Network, P.O. Box 8007, Green Bay, WI 54308; www.hyperacusis.net

Hyperacusis: Treatment

“There have been only a few studies outlining management for hyperacusis patients. Currently assessment indicates that the Jastrboff model for treating hyperacusis is the most widely accepted among practitioners. It is similar in purpose, to the Jastreboff approach for treating tinnitus: Tinnitus Retraining Therapy (TRT)...However, no one clinical approach has been sufficiently compelling to gain universal acceptance”. (p. 1 of 2)

Sandlin, R. (May 3, 2004). Current Research in Hyperacusis. Audiology On Line: From website: www.audiologyonline.com/askexpert/display_question.asp?question_id=224, p. 1 of 2.

TRT Therapy Developer

- ❖ Pawel J. Jastreboff, Ph.D., Sc.D., M.B.A.
- ❖ Professor and Director
- ❖ Tinnitus and Hyperacusis Center
- ❖ Emory University
- ❖ Atlanta, Georgia
- ❖ www.tinnitus-pjj.com



Hyperacusis Treatment in Asperger's Disorder

“It is important to first identify which auditory experiences are perceived as painfully intense, with the child communicating distress by covering his ears, flinching or blinking in response to sudden noises, or simply telling an adult which sounds are hurting” (p. 277).

Attwood, T. (2007). The Complete Guide to Asperger's Syndrome. Philadelphia, PA: Jessica Kingsley, p. 221.

Treating Hyperacusis in Those with Asperger's Disorder

- Remove the sound from the environment
- Use sound suppression (i.e., silicone ear plugs, sound suppression – Bose Quiet Comfort headphones, a fan, etc.
- Explaining the cause and the duration of the painful sound may be helpful: Carol Gray's "Social Stories" offer such information - <http://www.thegraycenter.org/>

Attwood, T. (2007). The Complete Guide to Asperger's Syndrome. Philadelphia, PA: Jessica Kingsley, p. 221.

Recruitment: Treatment

“New digital hearing aids are being developed which employ sound compression and volume control. These devices hold promise as technology improves however each person with recruitment is different and a good hearing aid would have to be customized to the patient’s recruited ears. In fact, the two ears on an individual with recruitment may have different levels of hearing loss, thus a pair of hearing aids may have to be tuned differently for each ear” (p. 4 of 25)

The Hyperacusis Network (No Date) Supplement. From Website:
www.hyperacusis.net/hyperacusis/supplement/default.asp.

Controversial Hyperacusis Treatment



- Auditory Integration Therapy (Tomatis)
 - Developed by Guy Bernard of France
 - Listen to specially modulated music for 10 hours
 - **MAY** help somewhat, but ***no research exists to indicate its helpful and it costs a lot.***

Attwood, T. (1998). Asperger's Syndrome: A Guide for Parents and Professionals. Philadelphia, PA: Jessica Kingdley.

- Some believe this allows the cortex to reorganize and strengthens the muscles in the ear, but no research proves this.

Silver, L. B. (2001). Theme Editor's Note. Perspectives, 27 (3), 5.

Sinha, Y., Silove, N., Wheeler, D., Williams, K. (2004). Auditory Integration Training and other Sound Therapies For Autism Spectrum Disorders (Cochrane Review, Abstract). The Cochrane Library, 2, from: www.cochrane.org/cochrane/revabstr/AB003681.htm

Attwood, T. (2007). The Complete Guide to Asperger's Syndrome. Philadelphia, PA: Jessica Kingsley, p. 378-279.

Controversial Hyperacusis Treatment

- Sensory Integration Training:
 - “The therapy uses a range of specialized play equipment to improve the processing, modulation, organization and integration of sensory information” (p. 278)
 - “Despite the popularity of this treatment, there is remarkably little evidence of the efficacy of sensory integration therapy” (p. 278)

Attwood, T. (2007). The Complete Guide to Asperger's Syndrome. Philadelphia, PA: Jessica Kingsley.

Helpful Websites about Hyperacusis

- The Hyperacusis Network:

P.O. Box 8007

Green Bay, WI 54308

earhelp@yahoo.com

www.hyperacusis.net



- www.ldonline.org
- National Institute on Deafness and Other Communication Disorders: nidcdinfo@nidcd.nih.gov
- American Academy of Audiology: www.audiology.org
- American Speech-Language Hearing Association: www.asha.org

Helpful Websites about Hyperacusis (Continued)

- American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS): www.entnet.org
- National Coalition for Auditory Processing Disorders: www.ncapd.org
- American Tinnitus Association: www.ata.org



Classroom Acoustics



- The American Speech-Language Hearing Association (www.asha.org) has a position paper on this that includes a comprehensive bibliography: Paper number 37, supplement 14.
- The Counsel of Educational Facility Planners International (CEFPI) has the following article on there website about how to build in good classroom acoustics:
- Erdreich, J. (July, 1999). Teaching in the Dark. Brief on Educational Facilities.