(Central) Auditory Processing Disorder (CAPD) & Hyperacusis
2016 Update
Central Auditory Processing Disorder

- “The inability to understand spoken language in a meaningful way in the absence of what is commonly considered a hearing loss.” (Sineps and Hunter, 1997)
- “…auditory processing disorders can coexist with hearing loss…” (Bellis, 2002)

Auditory Processing

“Auditory processing is the ability to attend, discriminate and understand the spoken message, particularly in the presence of competing stimuli and adverse listening conditions.” (p. 2)

Central Auditory Processing

“Central auditory processing is the perceptual processing of auditory information in the central nervous system. It involves several mechanisms that underlie abilities such as discrimination, recognition, temporal integration, localization of the signal in the presence of competing conditions and under degraded acoustic signals.” (p. 2)

NIDCD Definition of (C)APD

“Auditory processing is a term used to describe what happens when your brain recognizes and interprets the sounds around you. Humans hear when energy that we recognize as sound travels through the ear and is changed into electrical information that can be interpreted by the brain. The ‘disorder’ part of auditory processing disorder means that something is adversely affecting the processing or interpretation of the information.” (p. 1 of 3)

Definition of (Central) Auditory Processing Disorder:
“(C)APD is a deficit in neural processing of auditory stimuli that is not due to higher order language, cognitive, or related factors. However, (C)APD may lead to or be associated with difficulties in higher order language, learning and communications functions. Although (C)APD may coexist with other disorders (e.g., attention deficit hyperactivity disorder [ADHD], language impairment, and learning disability) it is not the result of these disorders.” (p. 1 of 26)
American Speech-Language-Hearing Association Definition of (C)APD

“Broadly stated, (Central) Auditory Processing [(C)AP] refers to the efficiency and effectiveness by which the central nervous system (CNS) utilizes auditory information. Narrowly defined, (C)AP refers to the perceptual processing of auditory information in the CNS and the neurobiologic activity that underlies that processing and gives rise to electrophysiologic auditory potentials. (C)AP includes the auditory mechanisms that underlie the following abilities or skills: sound localization and lateralization; auditory discrimination; auditory pattern recognition; temporal aspects of audition, including temporal integration, temporal discrimination (e.g., temporal gap detection), temporal ordering, and temporal masking; auditory performance in competing acoustic signals (including dichotic listening); and auditory performance with degraded acoustic signals”.

Important Point

- CAPD can effect children, adolescents and adults

Symptoms of (C)APD

“Children with auditory processing disorders appear to be uncertain about what they hear, and may have difficulties listening when there is background noise, following oral instructions and understanding rapid or degraded speech in the presence of normal peripheral hearing.” (p. 361)

Symptoms of (C)APD

- Difficulty performing multi-step directions
- Poor listening skills
- Slow auditory processing speed
- Language problems – developing vocabulary and understanding the spoken word, etc.
- Problems with reading, verbal and reading comprehension, spelling and vocabulary
- Poor academic performance
Symptoms of (C)APD

- Behavior problems
- Problems remembering and recalling information presented orally
- Problems attending to auditory information

Behavioral Symptoms of CAPD

• “difficulty understanding speech in the presence of competing background noise or reverberant acoustic environments
• problems with the ability to recognize the source of a signal
• difficulty hearing on the phone
• difficulty following rapid speech
• difficulty or inability to detect the subtle changes in the prosody that underlie humor and sarcasm
Behavioral Symptoms of CAPD

• difficulty learning a foreign language or novel speech materials, especially technical language
• difficulty maintaining attention
• a tendency to be easily distracted
• poor singing, musical ability, and/or appreciation of music
• academic difficulties, including reading, spelling and/or learning problems.” (p. 5)

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Factors involved in (C)APD

Recently researchers have found two possible cognitive factors related to CAPD. One is “working memory and executive attention” and the other is “processing speed and alerting attention”. Such factors have been found to negatively effect reading and math skills development and be related to several neurobiological disorders. If an individual also has a general difficulty with auditory processing in addition to the above this may explain the high number of comorbidities with CAPD.

Other Thoughts

- CAPD may in some cases be related to neurological disorders the minority of the time, but it is important to know this.
- Some children may have CAPD because they are developing somewhat slower than their age peers in their central nervous system functions. They may catch up eventually to their non-impaired peers.
- Ectopias, hetrotopias (cells in the wrong place), and pyromicrogyria (Multiple underdeveloped gyri) in the brain may impact auditory function and processing.
- Those who have problems with mylenization would be expected to be at risk of CAPD due to signal degradation.
- Many with dyslexia have been found to have CAPD comorbidly. It may be some of the altered neurobiology of dyslexia is related to CAPD due to it’s primary symptom of weak phonemic awareness.

Other Thoughts

Children with CAPD have a similar weakness of auditory processing of the left ear during dichotic listening that is found in those whose corpus callosums have been cut by split brain surgery. This is assuming the child’s auditory processing dominate hemisphere is the left hemisphere. Similar left ear deficits have been found in those with specific learning disorder.

Neuroanatomy of the Auditory System

- **Auditory Nerve**: 8th cranial nerve – Connects the cochlea to the brainstem – It allows the passage of the information from the approximately 30,000 nerve fibers of the cochlea to the brain

- **Cochlear Nucleus**: First structure of the Central Auditory System translates physical sound waves into electroneural impulses

- **Superior Olivary Complex**: Part of the brainstem that first connects impulses from both cochleas – Part of the ascending and descending auditory system

- **Lateral Lemniscus**: This is a tract of neurons in the brainstem that carries sound information from the cochlea to the contralateral inferior colliculus of the midbrain
Neuroanatomy of the Auditory System

- **Inferior Colliculus**: Midbrain area that controls eye movements
- **Medial Geniculate Body**: Part of the auditory thalamus – responsible for the intensity, duration and detection of sound
- **Auditory Cortex**: Part of the temporal lobe that processes sound
- **Corpus Callosum**: Responsible for communication between the hemispheres of the brain

Central Auditory Processing Disorder

• CAPD is not well defined
• May be due to under myelinated neurons in the corpus callosum.
• Those with CAPD process sounds at a slower rate.
• High rate of Otitis Media (ear infections)

Causes of CAPD

“Perhaps the most obvious examples are cases of complete central deafness, in which individuals show pronounced auditory deficits due to lesions existing primarily in the brain, despite presence of normal peripheral systems.” (p. 5)

Causes of (C)APD

“In terms of pathophysiological mechanisms, APD may be classified as occurring in the presence of: neurological conditions; delayed central nervous system maturation; or other developmental disorders.” (p. 362)

Causes of (C)APD

“Although most individuals with (C)APD do not exhibit frank lesions of the CANS (Central Auditory Nervous System), there is substantial evidence that many individuals with (C)APD do, upon autopsy, exhibit neuromorphological abnormalities in auditory areas of the CNS (Central Nervous System). Moreover, the same or similar patterns of test findings that are seen in anatomically confirmed central auditory dysfunction also appear in children and adults suspected of having (C)APD who exhibit no frank lesion or pathology.” (p. 10 of 26)

Causes of (C)APD

- Tumors of the Central Auditory Nervous System (CANS)
- Prematurity/Low Birth Weight
- Brain Damage
  - Meningitis
  - Head Trauma
  - Heavy Metal Poisoning
  - Lyme Disease
- Cerebrovascular Disorders
- Metabolic Disorders
- Epilepsy
- Delayed Maturation of the Auditory System

Diagnosing CAPD

- Team approach:
  - Audiologist (Case Manager)
  - Speech-Language Pathologist
  - Educator
  - Psychologist
  - Parents


**National Coalition for Auditory Processing Disorders**: [www.ncapd.org](http://www.ncapd.org)
What is Needed in a (C)APD Assessment

- Occupational Therapist: Evaluates sensory systems
- Speech-Language Pathologist: Language assessment
- Psychologist: Neuropsychological and cognitive/emotional assessment
- Audiologist: Auditory evaluation


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Kevin T. Blake, Ph.D., P.L.C.
CAPD Diagnosis

“Administration of behavioral and/or electrophysiologic audiologic tests that have been shown to be sensitive and specific to dysfunction of the CANS is critical for a proper diagnosis of CAPD, in addition to assessments and collaboration with a multidisciplinary team.”

Diagnosis of CAPD

“The diagnosis of (C)APD should be made on the basis of a carefully selected battery of sensitive and specific behavioral tests and electrophysiologic procedures, supplemented by observation and detailed case history.” (p. 5)

Diagnosis of CAPD

“The diagnosis should be made by audiologists who have been properly trained in the area of (C)APD, including the administration and interpretation of these tests and procedures.” (p. 5)

Diagnosis of CAPD

• Evaluations for (C)APD should be done in a soundproof room with acoustic control of environment and test stimuli.

(C)APD and Peripheral Hearing Loss

• The ASHA working group on (C)APD stated that (C)APD can be diagnosed in individual’s with peripheral hearing loss.

• “The experienced audiologist can apply several strategies in administering and interpreting central auditory tests to minimize the degree to which peripheral hearing loss influences central auditory test interpretation.” (p 9 of 26)

(C)APD and Peripheral Hearing Loss

• (C)APD evaluations can be done on children, adolescents and adults.
• Such evaluations can even be done with those with peripheral hearing loss if special care and procedures are followed.

Diagnosing CAPD

• Physicians – “If there is a disease or disorder related to hearing, you may be referred to an otolaryngologist, a physician who specializes in diseases and disorders of the head and neck.” (National Institute on Deafness and Other Communication Disorders, May 8, 2002, p. 3)

• American Medical Association: www.ama-assn.org
ADHD Vs CAPD

• “It is often too difficult to differentially diagnose the two, particularly since the rate of co-occurrence is so high.” (p. 2)
• 41 to 83% of children with CAPD have comorbid ADHD.
• It is not yet known what percentage of ADHD children have CAPD.

CAPD and AD/HD

• Comorbidity rates between 45 and 75%
• CAPD will often respond to stimulant medication. (Tannock and Brown, 2000)
• Audiologist Vs Psychologist/Psychiatrist:
  – CAPD or AD/HD?
• CAPD may be Inattentive AD/HD (Barkley, 2002)


“The relationship of ADHD to the language processing problem known as central auditory processing disorder (CAPD) is uncertain. Some researchers imply that they may not be separate disorders at all, given that teacher ratings of inattention in children with ADHD were significantly related to several tests of auditory processing. The problem here is largely though not entirely due to problems in definition.” (p. 131)

“Children with ADHD often have difficulty with auditory vigilance or attention…, and so they may automatically qualify for a diagnosis of CAPD on that basis alone.” (p. 131)

“There are many disorders that can affect a person's ability to understand auditory information. For example, individuals with Attention Deficit/Hyperactivity Disorder (ADHD) may well be poor listeners and have difficulty understanding or remembering verbal information; however, their actual neural processing of auditory input in the CNS is intact. Instead, it is the attention deficit that is impeding their ability to access or use the auditory information that is coming in”.

## Symptom Differences

<table>
<thead>
<tr>
<th>AD/HD, CT</th>
<th>(C)APD</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Inattentive*</td>
<td>1. Problems hearing noise</td>
</tr>
<tr>
<td>2. Distracted*</td>
<td>2. Problems following oral instructions</td>
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<tr>
<td>3. Hyperactive</td>
<td>3. Poor listening skills</td>
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<tr>
<td>4. Restless or Fidgety</td>
<td>4. Academic problems</td>
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<tr>
<td>5. Impulsive</td>
<td>5. Poor Auditory Association Skills</td>
</tr>
<tr>
<td>6. Butts in/Interrupts</td>
<td>6. Distracted/Inattentive*</td>
</tr>
</tbody>
</table>
“What is clear...is that CAPD and ADHD are not identical disorders if more rigorous definitions and criteria are used to determine the presence of CAPD, apart from merely clinical complaints of auditory inattentiveness. It remains uncertain whether CAPD should be considered a valid disorder apart from other already well-documented language disorders of children or whether it merely represents a more recent relabeling of those previously identified language disorders” (p. 131)

Barkley on AD/HD Vs CAPD

• Barkley indicated the following:
  • Some studies have found that those with CADP have an improvement in relief from their inattentive symptoms when they are administered stimulant medication. He believes such individuals have co-morbid ADHD and the stimulant medication is reducing their inattentiveness.
  • He believes 33% of those with ADHD also have CAPD. The general population’s rate is 3 to 5%.

AD/HD and Central Auditory Processing Disorder

• Tannock and Brown reported 45% to 75% comorbidity between AD/HD and CAPD.

• Hynd reported 50% of those with CAPD have AD/HD and 87% of those have comorbid Learning Disorders.


AD/HD and Central Auditory Processing Disorder

• What appears to be comorbid CAPD in those with AD/HD may be a problem with inhibition and subsequent distraction leading to uncertainty of what was heard.

• There may only be symptom overlap with CAPD.

The Development of Auditory Processing in Autistic Children.

Japanese researchers found children with autism are significantly delayed in the development of their auditory processing system when compared to non-disabled children. This they believe causes the language difficulties typical in autistic children.

Auditory Processing Disorder and Autism Spectrum Disorder

“...children with autism may have great difficulty with spoken language comprehension. However, it is the higher-order, global deficit known as autism that is the cause of their difficulties, not a specific auditory dysfunction”.

Sensory Processing in Those with ASD

British researchers reported results of a study that indicated that those children with ASD significantly different audiovisual, auditory, and visual processing of social facial, and speech stimuli than typically developing children. Those with ASD have significantly less activation in the brain areas that do the above processing and in the frontal lobe when exposed to social stimuli.

K.D. Tsatanis (2014) stated the problems with auditory processing often seen in those with ASD appear to be a part of their social world from infancy and can cause them difficulty in understanding social interactions as well as planning and coordination appropriate social responses.

Auditory Habituation in ASD Adults

Scientists found that adults with ASD are significantly slower than non-ASD adults at habituating to simple stimuli. They remained aroused loner after being exposed to sounds and they have a prolonged period of fear extinction of stimuli that is much longer. The researchers speculated this may explain why exposure therapy often takes longer in adults with ASD.

"Intervention recommendations for CAPD diagnosis are based on the demonstrated auditory processing deficits and related listening and related complaints".

Treating CAPD

- Help with Grieving Process
- Environmental Modifications
  - FM Loop Systems, Amplification, Seating, Etc.
- Remediation (Direct Therapy)
  - Phonological Awareness, Temporal Patterning, Prosody Training, Interhemispheric Training
- Compensatory Strategies

Recently top researchers in the field of CAPD stated that although the efficacy findings for auditory training for CAPD is somewhat limited current research support the use of auditory training.

Dichotic Interaural Intensity Difference (DIID)

Children with CAPD have been found to have weaker auditory processing in their left ears (assuming their left brain hemisphere is their language dominate one) during dichotic listening that would be expected. When trained with dichotic listening with the intensity raised to the weak ear some children are able to raise the level of that ear to the level of their “strong ear”. This technique may also work with people with brain injuries that suffer similar weak ears as a result of this accident.

Other Possible Training Approaches

- Earobics (www.earobics.com)
- Fast ForWord (www.fastforword.com)
- Lindamood-Bell Programs (www.lindamoodbell.com)

Discuss these with an audiologist from a university speech and hearing center.
**FM Loop System**

Website: [www.harriscomm.com](http://www.harriscomm.com/catalog/default.php?cPath=1141_46_158)
Sound Suppression Technology

Bose QuietComfort Sound suppression headphones:
www.bose.com
How to Make your Classroom Acoustically Available

• CAPD students sit in first row.
• Provide good lighting in the room.
• Avoid assigning a teacher to student who DOES NOT speak with a common or local accent.
• Acoustical tile in the ceiling
• Carpeting with thick carpeting pad on the floor
• Beards and moustaches need to be well trimmed away from lips. This allows for better speech reading.
• No mini-blinds! Draperies! Draperies absorb ambient sound better.
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 their website about how to build in good classroom acoustics:

  Brief on Educational Facilities.
Hyperacusis
Early Hyperacusis Definition in US

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

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)

“Hyperacusis is a disorder of loudness perception, in which sound intensities that are considered comfortable by most people are perceived unbearably loud.”

About 15% of the population has been estimated to have hyperacusis.

The American Academy of Otolaryngology Head and Neck Surgery defines Hyperacusis as:

“Hyperacusis is a condition that arises from a problem in the way the brain's central auditory processing center perceives noise. It can often lead to pain and discomfort.”

Author (No Date). Hyperacusis: An Increased Sensitivity to Everyday Sounds. From website: 
http://www.entnet.org/content/hyperacusis-increased-sensitivity-everyday-sounds.
Other Definition of Hyperacusis

The American Speech-Hearing Association Defines Hyperacusis as:

Hyperacusis is a rare hearing disorder that causes sounds which would otherwise seem normal to most people to sound unbearably loud.

Other Definition of Hyperacusis

The Hyperacusis Network describes the people who have the disorder as having, “…a collapsed tolerance to normal environmental sound. The term commonly used to describe this condition is 'hyperacusis'. Hyperacusis can come on gradually or occur suddenly where the patient finds themself in a state of crisis. Patients who have a collapsed tolerance to sound need to have their Loudness Discomfort Levels (LDL's) established by a hearing healthcare professional. Normal LDL's are in the 85-90+ decibel range. Patients with hyperacusis would have LDL's well below that level.

Hyperacusis is often associated with “piercing or burning” pain. It is often associated with low-intensity sound. The neurobiology of this pain is similar to other types of pain.

Similar Disorders
Hyperacusis

• There are four types of these extreme sensitivities to sound:
  – “Hyperacusis” which can affect those of all ages and is usually associated with tinnitus. They have hearing losses.

The Hyperacusis Network (No Date). Hyperacusis: Available from: 444 Edgewood Drive, Green Bay, WI 54302; web- dmalcore@mail.wiscnet.net

  – “Painful Hearing” in which certain frequencies are perceived as painful. Those with Developmental Disorders often have these problems. Approximately 40% of those with Autism and 95% of those with William’s Syndrome have this.

Hyperacusis-Types (Continued)

• “Super Hearing” occurs when individuals who can hear sounds extremely well, but it’s not physically painful.


• “Recruitment” these are people who have hyperacusis symptoms but have no hearing loss. This must be documented by an audiogram.

   The Hyperacusis Network (No Date). Hyperacusis: Available from: 444 Edgewood Drive, Green Bay, WI 54302; web- dmalcore@mail.wiscnet.net
Recruitment: “...is the rapid growth of perceived loudness for sounds in the pitch region of a person who has hearing loss. 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."

“HYPERSENSITIVE HEARING (OF SPECIFIC FREQUENCIES): “…are individuals who are sound sensitive at birth but it is only specific to certain frequencies heard at loud levels (typically above 70 decibels). It may seem like we are splitting hairs here but remember – the key words with hypersensitive hearing are – sound sensitive to specific frequencies heard at loud levels. These frequencies are typically labeled 'problem' frequencies. Autistic children are good examples of this. They can tolerate some sound at normal or even loud volumes but some frequencies are difficult to tolerable.”

“Misophonia (dislike of sound) is also an adverse response to sound no matter what volume the sound is. Typically misophonia characterizes an individual who reacts strongly to soft sounds and sometimes is further triggered by seeing the source of the offending sound. Common examples would include but are not limited to: the sound of people eating, smacking their lips, sniffing, the sound of certain consonants like p, s, or t, and repetitive sounds.” There is a significant percentage of these people who have obsessive-compulsive disorder.

“Phonophobia is considered a subcategory of misophonia. This is also an adverse emotional response involving fear of sound. Phonophobia often develops with an individual who has a significant collapsed tolerance to sound. They not only fear the sound of the environment they are experiencing in real time (right now) they worry about the sound that future events of the day or in the near future will produce.”

Hyperacusis Facts
Facts About Hyperacusis

- Loud and high frequency sounds can be very troublesome
- Ambient noise can be difficult, too
- This can cause social isolation, depression, and phonophobia, or the fear of sounds
- It is estimated 50,000 Americans suffer from hyperacusis.
- It can effect one, or both ears
- It effects children, adolescents and adults

Author (No Date). Hyperacusis: An Increased Sensitivity to Everyday Sounds. From website: [http://www.entnet.org/content/hyperacusis-increased-sensitivity-everyday-sounds](http://www.entnet.org/content/hyperacusis-increased-sensitivity-everyday-sounds).
Possible Causes
Possible Causes of Hyperacusis

- Temporomandibular joint (TMJ) syndrome
- Bells Palsy
- Lyme disease
- Viral ear infections
- Medications/toxins that damage the ear
- Airbag deployment

Author (No Date). Hyperacusis: An Increased Sensitivity to Everyday Sounds. From website: http://www.entnet.org/content/hyperacusis-increased-sensitivity-everyday-sounds.
Mechanism?

“This strongly suggests that the central auditory system plays a role in the development of hyperacusis. The central auditory system is the complex and adaptive network of neurological pathways from the cochlea to the higher levels of the brain. There are two key features of this system that those with hyperacusis should know,

- The brain is highly adaptive and will adjust loudness and processing according to average sound exposure, even at the lowest levels of the brainstem. Adaptivity can lead to hyperactivity when cells are damaged in the cochlea.

- It is thought that hyperactivity without sound stimulation may be an expression of tinnitus and sound-induced hyperactivity may be an expression of hyperacusis.”

American scientists found special nerves that communicate with information from the cochlea to the brain stem that warns the person of sounds that can damage the ears. This system only activates when there is damaging sound detected. This system is call auditory nociception. Type I auditory neurons transmit meaningful sound, and Type II auditory neurons alert to damaging sound.

Possible Causes of Hyperacusis

- Tay-Sack’s Disease
- Chronic Fatigue
- Post Traumatic Stress Disorder (PTSD)
- Some types of Epilepsy
- Migraines
- Depression
- The medication Valium

- Autism Spectrum Disorder (possibly 40% of those with ASD have Hyperacusis)
- Cerebral Palsy

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Asperger’s Disorder and Hyperacusis

- 70 to 80% of those with Autism Spectrum Disorder have some form of Hyperacusis.

- The three types of sounds people with Autism Spectrum 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.

A literature review published in 2007 found that perhaps 90% of those with autism spectrum disorder have comorbid hyperacusis. There is no known reason why this it, but it may indicate the individual have an overall sensory hypersensitivity.

American scientists discovered through using evoked potential with adults with autism they had more sensitivity to auditory stimuli than non-autistic adults.

Other Causes

Other possible causes:

Exposure to loud noise without proper ear protection

Endocrine problems: Addison's disease & Hyperthyroidism


http://www.asha.org/uploadedFiles/AIS-Hyperacusis.pdf#search=%22Hyperacusis%22.

Author (2016). *Sound Therapy & Counseling. Hyperacusis Focus*. From website:

http://hyperacusisfocus.org/research/causes/.
Others At Risk For Hyperacusis

- **Musicians**

- **Call center workers**


Comorbidities
Comorbidities to Hyperacusis

Hyperacusis can be associated with hearing loss, tinnitus, Lyme disease, William’s syndrome, and head/neck injuries. Hyperacusis has been found to be genetically connected to William’s Syndrome. Hyperacusis may be connected to sensitivity to strong smells, headaches, problems with bright lights and balance problems. Hyperacusis in some may be connected to chronic inflammation to the cochlea.


“Williams Syndrome is a genetic condition that is present at birth and can affect anyone. It is characterized by medical problems, including cardiovascular disease, developmental delays and learning disabilities. These occur side by side with striking verbal abilities, highly social personalities and an affinity for music... Individuals with Williams Syndrome have a very endearing personality. They have a unique strength in their expressive language skills and are extremely polite. They are typically unafraid of strangers and show a greater interest in contact with adults than with their peers.”

Williams Syndrome is a rare condition caused by missing genes. Parents may not have any family history of the condition. However, a person with Williams syndrome has a 50% chance of passing the disorder on to each of his or her children. The cause usually occurs randomly.

Williams Syndrome occurs in about 1 in 8,000 births.

One of the 25 missing genes is the gene that produces elastin, a protein that allows blood vessels and other tissues in the body to stretch. It is likely that having only one copy of this gene results in the narrowing of blood vessels seen in this condition.

Williams Syndrome
(WEBSITE: 46)

Those with Williams Syndrome have brains that are 20% smaller, particularly in the back of the brain which includes the occipital and parietal lobes. The temporal lobes are either normal in size or larger than normal. Their planum temporale is larger than normal and hence they often have perfect pitch. Finally, they use their cerebellum, brain stem and amygdala to process music thus it provides them an uniquely emotional experience when they hear it.

ASD and Williams Syndrome

(WEBSITE: 47)

ASD and Williams Syndrome, “two sides of the same coin,” Allan Reiss Stanford Medical School professor said. Social behavior and communication are underdeveloped in ASD and overdeveloped in Williams Syndrome.

Hyperacusis has a high comorbidity with autism spectrum disorder (ASD). The CD 38 gene encodes partially with the oxytocin system and hormone which has been shown to be deficient in people with ASD, and has been shown to cause difficulties in auditory processing of social sounds, facial emotion recognition, and social behavior. Administering oxytocin has been found to help these and may help with hyperacusis.

Hyperacusis

- People with Asperger’s Disorder report the following types of Hyperacusis:
  - Sharp unexpected noises
  - High pitched continuous noise...i.e. small electric motors, etc.
  - Confusing complex or multiple sounds-shopping malls

Hyperacusis

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

From website:http://www.add.org/articles/hypersen.html
Tinnitus and Hyperacausis

- Hyperacausis can also be related to tinnitus, or ringing in the ears
- The American Academy of Otolaryngology Head and Neck Surgery estimates:
  - 36,000,000 Americans suffer from tinnitus
  - One out of 1000 people who have tinnitus also has hyperacausis

Author (No Date). Hyperacausis: An Increased Sensitivity to Everyday Sounds. From website: http://www.entnet.org/content/hyperacausis-increased-sensitivity-everyday-sounds.
More About Tinnitus

➢ “...tinnitus or head noises, which is the perception of sound without an external source being present...Tinnitus is commonly defined as hearing a sound in the absence of external sounds.”

➢ About one in 5 have the type of tinnitus that causes them persistent distress, health, and quality of life issues

➢ If it is caused by specific known problems in the hearing system it is called “secondary tinnitus”

➢ If the cause is not known it is called “primary tinnitus”

Author (No Date). Hyperacusis: An Increased Sensitivity to Everyday Sounds. From website: http://www.entnet.org/content/hyperacusis-increased-sensitivity-everyday-sounds.
More About Tinnitus

- Tinnitus can be caused by:
  - Hearing loss
  - Exposure to loud noise
  - Prescriptions medications
  - Over-the-counter medications
  - You can have it in one or both ears

Author (No Date). Hyperacusis: An Increased Sensitivity to Everyday Sounds. From website: http://www.entnet.org/content/hyperacusis-increased-sensitivity-everyday-sounds.
Other Causes of Tinnitus

- Too much caffeine
- Ménière's disease
- Head injury
- Hypertension
- Stress
- Anemia
- Smoking
- Wax in the ears
- Some tumors

“Tinnitus is a disorder of perception of phantom sound that is also known as ringing in the ear or head. Tinnitus affects 10–20% of the general population.”

“The prevalence of frequent tinnitus is highest among older adults, more common in men than in women, more likely in former smokers, and in adults with hypertension, hearing impairment, loud noise exposure, or generalized anxiety disorder.”

Comorbidity of Hyperacusis and Tinnitus

The American Tinnitus Association reported that 12% of its members with tinnitus reported having hyperacusis.


Others have reported that 30 to 40% of those with tinnitus have hyperacusis and that most people with hyperacusis have tinnitus.

Evaluating Hyperacusis
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)
  - a thorough ear examination
  - a thorough hearing examination
  - a thorough hyperacusis examination that includes a loudness discomfort test.

Diagnosing Hyperacusis

The American Academy of Otolaryngology Head and Neck Surgery states the following is involved in an evaluation for hyperacusis:

- A full audiological evaluation
- An evaluation by an Otolaryngologist (Ear, Nose, & Throat Doctor)
- A medical physical

Author (No Date). Hyperacusis: An Increased Sensitivity to Everyday Sounds. From website: http://www.entnet.org/content/hyperacusis-increased-sensitivity-everyday-sounds.
Hyperacusis Treatment in Autism Spectrum 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)

More About Hyperacusis Evaluation

- Types of Audiological exams needed:
  - Otoacoustic Emissions: Test of cochlear hair functions
  - Tinnitus Matching: If one has tinnitus comorbidly this test can determine what the person’s tinnitus sounds like
  - Comprehensive Audiological Evaluation
  - Tympanometry: Test of pressure in the middle ear
  - Acoustic Reflex Thresholds: Test of middle ear and auditory nerve

German researchers found the Tinnitus Sample Case History Questionnaire (TSCHQ-
http://www.tinnitusresearch.org/en/consensus/consensusdocuments/en/TINNITUS_SAMPLE_CASE_HISTORY_QUESTIONNAIRE.pdf) can be used as a good screener for hyperacusis because of its focus on pain and fear related to sound/hyperacusis.

English Language Questionnaire for Hyperacusis

Sensory Sensitivity Symptoms in Autism Spectrum Disorder


Treating Hyperacusis
Hyperacusis Treatments

- Ear plugs, sound suppression head phones/ear buds, ear muffs

**BUT**

- If someone does this it can cause them to be more sensitive to sounds after they remove the ear protection

- One option is 6 months to a year of sound desensitization therapy
  - Some believe that Auditory Integration Therapy (AIT) may be helpful, but the Association of Speech-Language Pathologists (ASHA) does not believe there is sufficient scientific research to indicate it works.

- What may work is biofeedback, relaxation training, and acupuncture


Hyperacusis Treatments

Wearing ear protection constantly may make the brain even more sensitive to sound and may backfire. The individual with hyperacusis should be encouraged to expose themselves slowly to more difficult to tolerate sounds so they can be desensitized to them. Counseling may help with this, too.

Hyperacusis Treatments

Relaxation training and hypnosis for anxiety control may be helpful, too.

Hyperacusis Treatments

The American Academy of Otolaryngology Head and Neck Surgery states on its website:

- These is no specific medical treatment
- Sound therapy using a white noise generation may be helpful

Author (No Date). Hyperacusis: An Increased Sensitivity to Everyday Sounds. From website: [http://www.entnet.org/content/hyperacusis-increased-sensitivity-everyday-sounds](http://www.entnet.org/content/hyperacusis-increased-sensitivity-everyday-sounds).
Hyperacusis: Treatment

“There have been only a few studies outlining management for hyperacusis patients. Currently assessment indicates that the Jastreboff 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)

Hyperacusis Treatments

“The common treatment for hyperacusis is listening to broadband pink noise though sound generators (special hearing aids) which must be ordered through a specially trained doctor or audiologist who administers Hyperacusis (Tinnitus) Retraining Therapy.” This type of train takes about 6 months. People trained by Pawel J. Jastreboff, Ph.D., Sc.D., M.B.A, a professor at Emory University in Atlanta, GE, know this treatment (http://www.tinnitus-pjj.com/referral.html). The Hyperacusis Network sells CD with pink noise on them also (www.hyperacusis.net).

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
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
Treatment of Hyperacusis

- There are no cures
- Counseling and Sound Therapy with machines that expose the patient to low level background sounds help some.
- Neuropathic pain appears similar to auditory nociceptive pain and responds to medications that produce more of the neurotransmitter GABA.
  - The medications gabapentin (Neurontin®) and pregabalin (Lyrica®) have been found to help some with hyperacusis (first line treatment, works with 30% about 30% relief).
  - SSRI anti-depressants may also be helpful to some.
Treating Hyperacusis in Those with Autism Spectrum 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/social-stories


Cognitive Behavioural Therapy (CBT): “The idea behind using CBT is to recognise what is helpful and/or unhelpful in your everyday life when it comes to living with hyperacusis. You can then work with your therapist to find more helpful ways of managing your hyperacusis, thereby reducing the impact it has on you.”


Surgery for Hyperacusis

Those that have a tear, or defect in a perilymph fistula, membranes that separate the fluid of the inner ear from the air of the middle ear, and those who may have an acoustic neuroma, a benign tumor on the auditory nerve(s) that cause symptoms of hyperacusis can have surgery to correct these defects and growths.

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.*


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


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)

Possible Future Treatment

Low-level Laser Cochlear Therapy reportedly has been shown in 60 case studies to reduce hyperacusis symptoms. Two wavelengths, one in the visible red spectrum and the other in the inferred spectrum may be useful in the future to treat hyperacusis. The light may promote better circulation to damaged cochlear tissue and better cellular energy.

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.”

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

- The Hyperacusis Network: 
P.O. Box 8007
Green Bay, WI 54308
www.hyperacusis.net
- www.earhelp.co.uk
- Hyperacusis Focus: http://hyperacusisfocus.org/
- National Institute on Deafness and Other Communication Disorders: www.nicdinfo.nidcd.nih.gov
- American Academy of Audiology: www.audiology.org
- American Speech-Language Hearing Association: www.asha.org
Helpful Websites about Hyperacusis (Continued)

- National Coalition for Auditory Processing Disorders: [www.ncapd.org](http://www.ncapd.org)
- American Tinnitus Association: [www.ata.org](http://www.ata.org)
- British Tinnitus Association: [http://www.tinnitus.org.uk/contact-us](http://www.tinnitus.org.uk/contact-us)
Good Books On Hyperacusis


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 their website about how to build in good classroom acoustics: