HANDOUT ON THE MANAGEMENT OF HYPERACUSIS

(REGISTER NO, M 0104)

An Independent project submitted in part fulfillment of the First year M.Sc (Speech and Hearing), University of Mysore, Mysore

ALL INDIA INSTITUTE OF SPEECH AND HEARING
MANASAGANGOTHRI, MYSORE - 570006

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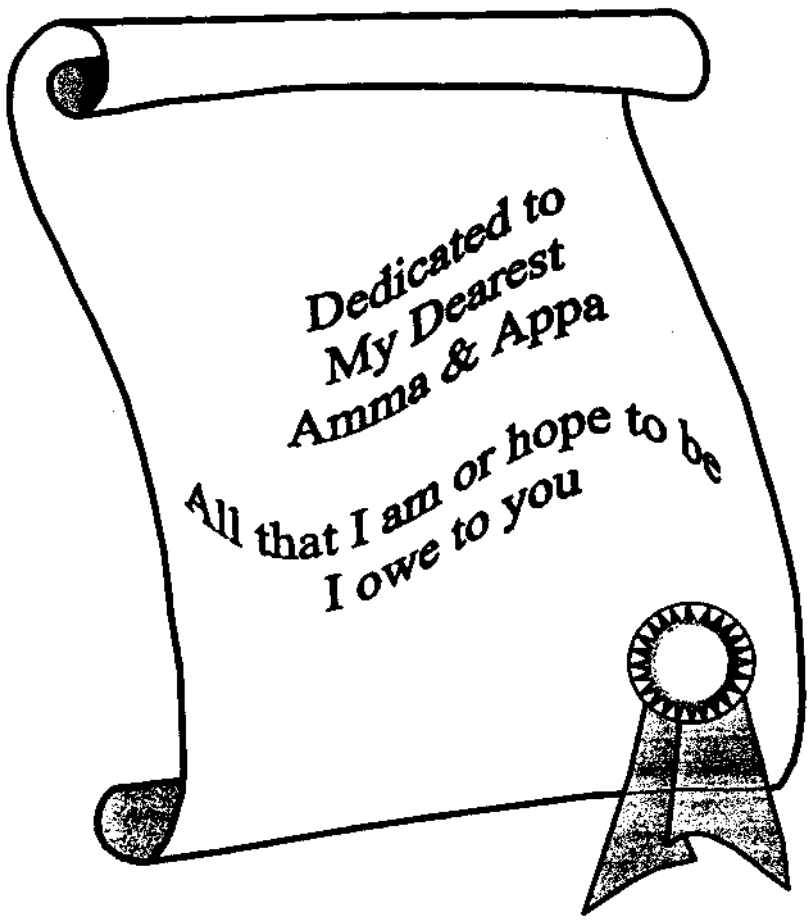
Dedicated to
My Dearest
Amma & Appa
All that I am or hope to be
I owe to you

Certificate

This is to certify that the Independent project entitled *"Handout On The Management of Hyperacusis* " is the bonafide work done in part fulfillment of the degree of Master of Science (Speech and Hearing) of the student (Register No. M 0104).

Mysore
May 2002

Director
All India Institute of
Speech & Hearing
Mysore - 570006.

Certificate

This is to certify that the Independent project entitled *"Handout On The Management of Hyperacusis* " has been prepared under my supervision and guidance. It is also certified that this has not been submitted earlier in any other University for the award of any Diploma or Degree.

Guide

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May 2002

Declaration

I hereby declare that this Independent project entitled *"Handout On The Management of Hyperacusis* " is the result of my own study under the guidance of Dr. K. Rajalakshmi, Lecturer in audiology, Department of audiology, All India Institute of Speech and Hearing, Mysore and has not been submitted earlier or in any other University for the award of any Diploma or Degree.

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- *Appa and Amma – I am grateful to God for having given me the world's best parents. Without you, I would have been nowhere.*
- *Shiv- My most troublesome brother. Every hour, every minute, every second, you have always been there for me. You have tolerated my tantrums, but I know you are my sweet brother, Dei- I adore you.*
- *My Dearest Roomie – Lucky to have a roommate like you. Thanks for everything else. Especially thanks for putting up with me.*
- *Mili- Thank you for being there for me anytime and every time.*

- *Ramya you are my right choice baby aaha..... Thanks for everything da.*
- *Vimi and Delcy- my philosopher and Guide in College life. Without you guys.....*
- *Tyagi- my daddy- your timely advices and suggestions and your moral support has made me grow up so far in college life.*
- *Mukunthan – My short-tempered brother, you will always hold a special place in my heart inspite of everything that has taken place.*
- *Prasanna and Anu- you have been two wonderful people and thank you for being there for me.*
- *Prakash- Though away, you have been a moral support to me. Thank you for the faith in me.*
- *Reddy, Divya madam and Sapna- thank you for your valuable suggestions and moral support.*
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- *All my seniors and juniors- thank you*

- *My dear classmates- thanks you guys.We r a wholesome bunch.love to be together.*
- *I would like to thank the librarians for their help.*
- *I would like to thank sir for his neat typing and a complete TP.*
- *I would like to thank God for giving me the strength to finish my project successfully.*

THE SILENT ONE

WE SILENT ONES DONT WANT TO HEAR, SOUNDS ONCE LOVED

THAT HURT THE EAR.

OUR LOVFED ONES VOICES ONCE SO DEAR FALL PAINFULLY

UPON THE EAR.

THE R0LLING WAVFES, THE OCEAN'S ROAR WE CANT ENJOY

THEM LIKE BEFORE,

THE SOUNDS OF LIVING, SOUNDS OF LIFE NOW BRING US

SUFFERINE AND STRIFE,

BUT SOME DAY THIS CRUEL ENEMY THAT ROBS LIFE'S

JOY FROM YOU AND ME.

WILL CONQUERED BE, AND ALL AROUND AGAIN WE WILL

JOY IN NATURE'S SOUNDS,

SO 'SILENT ONES' UNTIL THIS DAY'HOPE IN GOD'

THANKS AND PRAY.

BY HELEN GLEASON.

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INTRODUCTION

Hearing ability is a gift to mankind . Man is sensitive to a range of frequency of sounds. The frequency that a nonnal human ear can hear ranges from 20 Hz to 20 KHz. Every individual perceives a sound at a particular level. The normal range as specified ranges from 10 to 15 dB. If an individual needs to be given a louder level to perceive the sound, then, likely the chances that the individual suffers a hearing loss. While doing the audiometric evaluation, one part of the procedure is to obtain the uncomfortable loudness level. This can be obtained by increasing the intensity level and various stimuli like noise, speech or tone can be given. Generally, for normal individuals, the uncomfortable level is greater than lOOdB.

But in certain individuals, th,ere is a condition where they show hypersensitivity to sounds. This condition is also known as hyperacusis dysacusis or oxylacusis.

So, hyperacusis, as defined, is a relatively rare condition in which, the patient, with or without a hearing loss, experience severe loudness discomfort to everyday environmental sound levels. These people do not show abnormal loudness growth but an abnormal discomfort for suprathreshold sounds (Barnes and Marriage, 1995).

Hyperacusis is a poorly understood disorder resulting in many theories of e.,o.ogy and prognosis. Hyperacusis can occur a.one or in conjunction with other disorders. It is said that for those with this condition, the brain loses much

of its dynamic range, and everyday sounds are unbearably loud. A sudden single burst of noise (Schwade, 1995), a head injury (ASHA, 1995) or surgery to the face or jaw (Barnes and Marriage, 1995) can result in hyperacusis. Hyperacusis is not typical of occupational noise exposure.

Hyperacusis is generally seen in both ears and may occur gradually and suddenly. There is some speculation that the efferent portion of the auditory nerve is selectively damaged while the hair cells to hear the sound remains intact.

The audiograms for hyperacusis sufferers are typically normal. They show normal sound thresholds but the sensitivity level is above normal. The comfort level for most of these people is below 100 decibels. People with hyperacusis can experience discomfort at 40 to 50 dB or lower (Schwade, 1995). The disorder may also be frequency specific (Schwade, 1995). Not all sounds of the same loudness (Number of decibels) cause discomfort, but only sounds within a certain range. Thus a small change of frequency may cause discomfort at low volume.

Barnes and Marriage proposed two types of hyperacusis

1. Peripheral hyperacusis
2. Central hyperacusis

Peripheral hyperacusis is when the ears built-in mechanism against loud or sharp sounds seems to have been turned off. Several authors have documented auditory impairment causing changes in peripheral hearing

mechanism - which give rise to sensitivity for loud sounds. Absence of acoustic reflexes, positive history of vestibular disorders, Meniere's disease or perilymph fistula account for peripheral hyperacusis. In perilymph fistula, changes in perilymph pressure may cause acoustic reflex reversal and audiosensitivity (Gordan, 1983). Hyperacusis is also an otological complication of Herpes Zoster (Adour, 1994) and craniomandibular disorders (Erlander and Rubinstein, 1991).

Another type proposed by Barnes and Marriage (1995) is the central hyperacusis. It results in an inability to tolerate specific but not necessarily loud sounds. Certain sound waves reaching the inner ear are then somehow over amplified or magnified on the way to the brain or by the brain. A global sensitivity may exist to explain central hyperacusis. Barnes and Marriage (1995) listed a few clinical conditions as co-occurring with central hyperacusis: migraine, depression, pyridoxine deficiency, musicogenic epilepsy, post-traumatic stress disorder and chronic or post-viral fatigue syndrome. Some manic individuals also report having a much sharper sense of hearing (American Psychiatric Association, 1995). Children who have autism or pervasive developmental disorder may also have hyperacusis (American speech language hearing association, 1995).

Diagnosis of hyperacusis, arising from peripheral auditory impairment, can be made by applying standard audiological test techniques which include acoustic reflex and Brainstem evoked response. Peripheral hyperacusis is

distinct both in symptom, history and causation from the proposed central hyperacusis.

Patients with hyperacusis may also be divided into those hypersensitive to the loudness of sounds with a decreased puretone uncomfortable loudness level and those hypersensitive to certain specific sounds irrespective of loudness levels and decreased uncomfortable levels to specific sounds.

With the available evaluative procedures, once the patient's condition has been obtained, the management needs to be concentrated on. The strategies available for the management include.

1. A treatment that has acquired a mixed support is the use of earplugs. Some have also recommended filtered ear attenuation (FEA) earplugs, ER-15, ER-25 attenuators. Filtered ear attenuation as reported have been used successfully in several cases of hyperacusis. They were seemingly ideal because they attenuate with good uniformity across the frequency bandwidth.
2. A few other treatment procedures for hyperacusis include biofeedback and relaxation techniques (American speech language and hearing association, 1995). But this treatment lacks scientific evidence.
3. The most commonly used strategy is the "desensitization" which is done by the use of noise generating instruments that are worn on both ears. These instruments produce a gentle static like sound (white noise). Another method of desensitizing is the use of pink noise through headphones continuously for 2 hours in a day. Desensitizing is a long

process and can vary from few months to few years. This treatment usually improves one's tolerance to sounds.

Need of the Study

Individuals with hyperacusis face the following problems :

1. These individuals often experience physical pain / discomfort to everyday sounds. They are adversely affected and do not enjoy normal life.
2. Sometimes, these individuals alter their environment to deal with the problem which can include avoiding loud sound activities, attempting to muffle or decrease sounds, use earplugs etc.
3. Hyperacusis can be devastating to the patients career relationships and peace of mind.

Keeping in mind the above problems, this study has been taken up.

Also, the management strategies for these individuals are seldom documented in a compiled form. They are scattered in different periodicals, monographs etc. Thus, it becomes imperative to put them in a collective form inorder that, the handout is user friendly to the practicing professionals and clients.

Aim of the Study

1. To prepare a handout for individuals with hyperacusis, on how to cope up with their problem.
2. To highlight the importance of counselling in these individuals.

METHODOLOGY

The handout is prepared based on the materials available from the following sources.

1. Monographs
2. Periodicals
3. Websites and other sources

Information from all the sources have been compiled to suit the purpose of the study.

REVIEW OF LITERATURE

According to Stedman's Medical Dictionary (cited in Moeller, 2000) hyperacusis(hyperacusia)is: —• an abnormal acuteness of hearing due to increased irritability of the sensory neural mechanism. In Borland's medical dictionary (26th edn.) (cited in Vemon, 1998) hyperacusis is defined as "an exceptionally acute sense of hearing, the hearing threshold being unusually low".

The term 'hyperacusis' is, strictly speaking, a misnomer for the symptom, as it describes hyperacute hearing thresholds, rather than suprathreshold hearing sensitivity (Dorland's Illustrated Medical Dictionary, 1988).

Hyperacusis literally means oversensitive hearing. Patients with hyperacusis manifest severe loudness discomfort to everyday sounds presented at normal intensities. Hyperacusis dolorosa has been suggested as a general term for patients having discomfort to moderately loud sound levels regardless of their hearing sensitivity (Mathisen, 1969).

Vernon (1998) defines it as "a collapse of loudness tolerance so that almost all sounds produce loudness discomfort, even though the actual sound intensity is well below that judged to be uncomfortable by others". A few others have used "phonophobia" synonymously with "hyperacusis". Both the terms are used to describe an unusual hypersensitivity or discomfort induced by exposure to sound.

Mathisen (1969) defines phonophobia as "an abnormal discomfort for suprathreshold sound that does not annoy healthy individuals". Jespen (1963) defines phonophobia as the discomfort from sounds less than about 65dBSPL when hearing is normal.

Phonophobia is also referred to as the fear of sounds. Individuals so afflicted are fearful of being exposed to sounds that have caused pain or discomfort.

Finally, hyperacusis is a term to describe uncomfortable or painful hypersensitivity to sounds. It does not imply super hearing or the ability to detect sounds softer than normal thresholds.

The prevalence of hyperacusis in the general population is unknown and is probably underrelated when occurring with other medically recognized symptoms like tinnitus, headache or depression. The prevalence depends heavily on how it is defined. A self-group started by and for persons with hyperacusis (The Hyperacusis Network) has estimated that one in 50,000 persons may suffer from some degree of suprathreshold hyperacusis. The American Tinnitus Association (ATA2) note that hyperacusis is more common among tinnitus sufferers, but states that it is still 2%. But the true prevalence may be underestimated because many hyperacusic patients with normal hearing report being turned away by hearing health care professionals that their condition is purely psychological (Arnold, 1978). Also, a questionnaire on a clinical population by Sanchez and Stephens (1997) found that 8 % of tinnitus

sufferers have hyperacusis. Studies suggest that 3 million people in United States have hyperacusis.

What is the cause of this hyperacusis ?

It can be said that the origin of hyperacusis is in the central auditory system. One-way of describing this problem is to state that central auditory system gain or amplification for sound stimulation is excessively high.

It is also possible that there is a disruption in the efferent or descending pathway of the auditory system function which could play a role in hyperacusis. That is, without the usual amount of inhibition or modulation_of the cochlear activity by the efferent system, the brain is over stimulated by the acoustic sensory stimulation.

One another underlying cause for hyperacusis could be a reduction in a brain chemical called Serotonin. Serotonin controls the amount of information arriving at the brain from the sense organs.

The other causes stated include a sudden single burst of noise (Schwade, 1995), a head injury (American Speech Language Hearing Association, 1995). This would suggest that physical damage to the feedback system between the ear and the brain could also underlie hearing over sensitivity. The secondary mechanism underlying long-term hearing hypersensitivity is a "fear-potentiated" startle reaction.

Other contributing causes are thought to be Temperomandibular syndrome (TMJ), William's syndrome, Bell's palsy, Meniere's disease,

Tay-sach's disease, Lyme disease, air bag deployment viral infections involving inner ear or VIII cranial nerve.

Also as many as 40% of all autistic children are sensitive to noise, their condition being termed hyperacute hearing. Hypothyroidism has also been associated with this disorder.

But with all these causes stated, there is no universal relationship between any one abnormality and hypersensitivity to sound.

As reported by the authors, Anari. M, Axelsson, A., Eliasson, A. and Magnusson, L. (1999)-

Aetiology of hypersensitivity to sound as suggested by history - out of 100 subjects.

Aetiology	**No. of Patients**
Sound elicited	
Music	**31**
Occupational	**13**
Other noise	**07**
Infections	
Meningitis	**01**
Zoster oticus	**01**
Post-Op. Otitis	**02**
Otitis chronic	**01**
Upper Respiratory Tract Infection	**01**
Borrelia	**01**
Vestibular Neuronitis	**10**
Traumatic	
Head concussion	**03**
"Whiplash injury	**02**
Surges acoustic neuronoma	**02**
Trauma of External Auditory Canal	**01**
Lightning stroke	**01**
Miscellaneous	
Sudden deafness	**03**

Salicylate intoxication	**02**
Metabolic Meniere	**01**
Psychological-mental	**05**
Hereditary hearing loss	**01**
Autoimmune disease	**01**
Excluded	**08**

The above listed are the causes update for hyperacusis.

Types of Hyperacusis

The types of hyperacusis is based on

> Brandy and Lynn (1995) classification

> Barnes and Marriage classification

Brandy and Lynn (1995) (cited in Shoup.A.G., and Roeser. R. 1, 2000) differentiate between 2 types of hyperacusis patients.

a) Threshold hyperacusis: was suggested for patients with better hearing than age related hearing sensitivity norms.

b) Suprathreshold hyperacusis: was suggested for patients having discomfort to sounds less than 65 dBSPL when hearing sensitivity was normal.

As by Barnes and Marriage, the classification includes

> Peripheral Hyperacusis

> Central Hyperacusis

a) Peripheral Hyperacusis

There are several documented auditory impairment causing changes in the peripheral hearing mechanism which give rise to sensitivity for loud sounds.

The best-documented report for this is related to the loss of acoustic or stapedial reflex (AR). The effect of acoustic reflex is to reduce energy transmission through the middle ear, primarily in the low frequency region. This mechanism affords some protection to the cochlea, from high levels of noise exposure. Although it is a simple arc, there is evidence that the acoustic reflex is under tonic facilitatory influences from higher centres in the brain (Mangham, 1984). Hyperacusis has been described following abolition of the stapedial reflex as seen in idiopathic (Bell's) facial nerve palsy (Adour, 1982), Ramsay Hunt syndrome (Wayman, D.M., Pham, H.N., Byl, F.M., and Adour, K.K., (1990), Myasthenia gravis (Laurian, N., Laurian, L., and Sandov, R. 1983). Hyperacusis can also occur with Meniere's disease (Cawthorne, 1948), and perilymph fistula (Fukaya and Nomura, 1988). Changes in perilymph pressure may cause acoustic reflex reversal and "audio sensitivity" as proposed by Gordan (1983). Alternatively, endolympharic fluid pressure variation may modify the microdynamics of the cochlea and the basilar papillae causing changes in the excitation patterns of outer hair cellls. In spite of excluding patients with absent acoustic reflexes, positive history of vestibular disorder or a diagnosis of Meniere's disease, there still exist a number of people who complain of the inability to tolerate specific, but not necessarily loud sounds. Auditory sensitivity" is recognized if a patient reports discomfort for sounds that would be acceptable to most normally hearing people. It has been

noted by Hazell and Sheldrake (1991) that sound intolerance may be part of a global sensitivity, including bright lights and tactile stimulation. The fact that auditory and visual oversensitivity occur together (Subcutaneous Sumatriptan International study group, 1991) would argue for a common central causation rather than peripheral site of lesion. Certain sound waves reaching the inner ear are somehow over amplified or magnified on the way to the brain or by the brain.

Barnes and Marriage (1995) listed the following conditions as co-occuring with central hyperacusis.

? Migraine, depression, Pyridoxine deficiency, Benzodiozapine dependence, musicogenic epilepsy, Tay-sach's disease, post-traumatic stress disorder, and chronic or post-viral fatigue syndrome. Children who have autism or pervasive developmental disorder may also have hyperacusis (American Speech-Language Hearing Association, 1995).

Manifestations by the patients

Hyperacusis will describe the sensation caused by particular sounds as "painful", "startling" or "anxiety producing". Ears with hyperacusis often feel painful during the early stages of hyperacusis and frequently feel pressure on the eardrum. Their frequent complaint is the fullness of the ear. These individuals are adversely affected and do not enjoy normal life. Sometimes, these individuals alter their environment and way of living to deal with the problem. This could include avoiding loud sound activities, attempt to muffle

or decrease sounds, use earplugs etc. The other things that are difficult for them to bear but may sound funny are sounds of coughing, sneezing, snoring, clapping, loud laughing. Driving vehicles on roads, watching movies in theatres are also difficult for them. Hyperacusis can be devastating to the patient's career relationships and peace of mind. An individual with this problem may have to leave his career if sounds in his working environment become intolerable. Individuals with this condition also report that anxiety or fatigue makes their sound intolerance even worse. They also say that specific sounds cause physical pain and nerve grating. Exposure to such sounds causes an increase in pulse rate and sweating.

These individuals find it difficult to get sleep and increase in stress levels is noticed. Some patients have reported that they perceive their own voice as uncomfortably loud.

Some also perceive a change in their voice i.e. talking softer, sounds hoarse or even whisper. The reasons attributed to change in voice include.

- They talk softly or become hoarse when their ears are bad or have incurred a recent noise injury.
- Their ears are sensitive in the early morning and late evening, which can affect the volume of their voice.
- They tend to speak softly so as to evoke a soft, low volume response from whomever they are speaking to.

Children with such a condition may try or fuss upon exposure to loud noises. They may respond negatively to loud sounds and parents need to be alert to the

responses of the child. Children may fall down, cry, cover their ears or scream loudly as a response to loud sounds.

Hyperacusis and Tinnitus

There is an increased recognition that tinnitus frequently co-occurs with hyperacusis and that there is a shared association between the mechanisms of some types of tinnitus and hyperacusis (Hazell and Sheldrake, 1991). Axelsson and Anari (1993) considered that the frequent co-occurrence of hyperacusis with tinnitus, hearing loss and sometimes distortion suggested a cochlear rather than retrocochlear pathology.

In contrast, Coles and Sood (1988) proposed that the generator site of phonophobia must be above the brainstem nuclei involved in the stapedial reflex arc, and was more likely to be at a cortical level rather than within the brainstem.

Jastreboff and Hazell (1993) described hyperacusis as a "manifestation of increased central gain" and in some cases, this may be the sole cause of tinnitus as seen in normal hearing subjects who are deprived of sound in an anechoic chamber (Heller and Bergmann, 1953).

Vernon and Meikle (1981) (cited in Tyler, R., 2000) believe that hyperacusis could be intimately linked to tinnitus. The hyperacusis survey conducted by the American Tinnitus Association revealed 112 patients having both tinnitus and hyperacusis. Of this number, 53% said that hyperacusis was more debilitating than the tinnitus. 25% said that each problem was equally

disturbing, 16% said that tinnitus was more debilitating and 6% were uncertain. Many patients with hyperacusis also reported of some temperomandibular joint problem. Fullness of ear was reported by 83% of 112 persons with hyperacusis.

Assessment of Hyperacusis

A very comprehensive basic audiologic assessment is essential for starters (Hall and Mueller, 1998). The test battery should include pure tone audiometry and otoacoustic emissions (Transient evoked otoacoustic emissions and distortion product otoacoustic emissions) to rule out sensory hearing impairment, some measure of word recognition and also tympanometry to assess middle ear status.

As a rule, acoustic reflexes should not be recorded as the required intensity levels (80dB and greater) may cause the hyperacusic patient undue discomfort, distress or pain. Hyperacusis should be evaluated carefully by both direct frequency specific measurements of loudness discomfort and by a detailed assessment to detect any intolerance of specific environmental sounds. Loudness discomfort levels (LDL's) for pure tone signals of lOOOHz and higher are often helpful in documenting decreased tolerance to sound. When loudness tolerance is measured in hyperacusic patients, it is usually found to be most abnormal in the higher frequency (Vernon, 1992). Measurements of hyperacusis performed during subsequent visits provide an important indication of the progress of treatment (Hazell and Sheldrake, 1992).

Audiograms generally in these individuals show sensitivity at minus decibel levels. Because of this, hyperacusis patients may feel as though, in

addition to having less tolerance to sound, they can hear even better than those with normal ears.

Johnson, M. (1999) developed a tool for measuring hyperacusis. It was called the Johnson Hyperacusis Dynamic Range Quotient (JHQ). This procedure is recommended only for hyperacusis patients who have essentially normal hearing thresholds. It is not appropriate for patients with significant hearing loss who may suffer from recruitment or a combination of recruitment and hyperacusis. The author has given a hyperacusis rating scale.

Rating	**JHQ Range**	Comments
Mild	75-90dB	Nearly within normal limits
Moderate	50-74dB	Use ascending approach
Severe	30-49dB	Careful with tympanometry
Profound	0-29dB	Careful with voice/equipment

As of a study by Anari, M., Axelsson, A., Eliasson, A., and Magnusson, L. (1999) audiological tests included?

- Pure tone thresholds
- Speech recognition scores
- Acoustic reflex thresholds
- Tone Decay test
- SISI- Short Increment Sensitivity Index
- Distortion measurement
- Uncomfortable loudness levels for pure tones
- Uncomfortable loudness levels for specific tones.

- Pure tone Thresholds : Mean hearing level revealed a moderate hearing loss at the high frequencies. Completely normal hearing was found in 39 patients, 18 with unilaterally normal hearing.
- Speech Recognition : The speech recognition score in background noise did not reveal any unexpected data and were generally within normal limits.
- Uncomfortable Loudness Levels (ULLs) : They varied from extreme hypersensitivity with ULLs between 35 and 45 dBHL to ULLs > 110 dBHL at all frequencies. There was no correlation with pure tone thresholds.
- Acoustic Reflex thresholds (AR) : The acoustic reflex thresholds were without exceptions within normal limits.
- Tone Decay - 2/63 patients were seen with a pathological decay (> 30dB).
- Short Increment sensitivity index - revealed no conclusive results.
- Uncomfortable loudness level for specific sounds - They were generally lower than the uncomfortable loudness levels for pure tones.
- Distortion - There was no significant difference in the uncomfortable loudness levels for patients with and without distortion. As reported by the authors, a test battery is required to confirm hyperacusis and not just pure tone thresholds and uncomfortable loudness levels.

Objective assessment of hyperacusis (as by Barnes and Marriage (1995) in their study).

Initial investigations on the audiological manifestations of hyperacusis have shown no consistent changes.

However, Butler (1993) found that post-auricular myogenic (PAM) response is significantly increased in amplitude in chronic fatigue syndrome (CFS) patients compared with matched controls. Additionally, when attention was diverted from the auditory stimulus, there was almost complete abolition of the post- auricular myogenic response in controls, but little change was seen in the chronic fatigue syndrome patients.

Collett, L., Roge, B., Descouens, D., Moron, P., Dirverdy, F. and Urgell, H. (1993) described a reduced contralateral suppressive effect in the evoked otoacoustic emissions of hyperacusic autistic patients. 40% of autistics are known to have high levels of 5-hydroxytryptamine(5-HT) (Schain and Freedman, 1961) and 40% have hyperacusis (Autism Research Review International Editorial, 1990). But there are no investigations if they were the same subject groups. The two techniques (Post-auricular myogenic response and contralaterally masked otoacoustic emissions) demonstrate that it is likely to be the central processing systems of the auditory function that will give insight in to the mechanism of central hyperacusis.

Management

The management as such can be divided into the following stages.

- Patient evaluation - helps in determining the category of treatment. This has been discussed earlier.
- A session of retraining counselling
- Fitting of instruments (noise generators or hearing aids) and or or additional instructions on how to enrich and use environmental sounds
- Follow-up visits

Intensive directive or educational counselling plays a very major role in hyperacusic patients. We should be able to acknowledge the seriousness and validity of his or her problem. Reassure them that the problem is not a serious one. Once the reassurance is given, an indepth discussion of the tests, anatomy and physiology of hearing mechanism and how it relates to hyperacusis should be carried out. Educating the patient helps to relieve the fears and concerns present in the individual.

Counselling needs to be part of the management program throughout.

Fitting of instruments vary differently. Of these, the aid that has received mixed support is the earplugs.

Basically, an obvious technique is to reduce the perceived level of the uncomfortable sounds. So patient would try to protect himself or herself from whatever sound he or she finds intolerable. This hearing protection might solve the patient's immediate difficulty.

But Vemon and Jastreboff, P. advice against the use of earplugs (Schwade, 1995). Earplugs deprive the auditory system of sound. Earplugs will only excarberate the underlying problem.

By continually keeping the ears protected from day-to-day sounds, patient is creating an artificial hearing loss. The pathological-neurological activity that causes abnormal increase in hearing sensitivity will compensate for the artificial loss by further enhancing the internal gain of the auditory system. As the subcortical brain continues to enhance the internal sensitivity of the hearing system, even greater and greater hearing protection is required. This may lead to patients requiring constant hearing protection, even when in quiet.

Hence it has been said that ear defenders need to be used only where there is a likelihood of exposure to high intensity sounds which would be bothersome which means to say use defenders only when necessary.

But it has also been said that, in the initial period of hyperacusis (i.e.) sudden onset, it is necessary to protect the ear when in loud surroundings as it is liable to further injury.

Also there is a recommended use of filtered ear attenuation earplugs-FEA ear plugs ER-15, ER-25 attenuators. FEAs have been used successfully in several cases of hyperacusis. They are seemingly ideal because they attenuate with very good uniformity across the frequency bandwidth.

Sound Desensitization

Sound therapy helps to realize the maximum tolerances left in the ears. The special sound therapy that has been divided to be administered on both tinnitus and hyperacusis patients is TRT-Tinnitus Retraining Therapy by Jastreboff, P. (1990).

Hyperacusis and tinnitus are two sides of the same coin and hence the treatment term TRT is applicable to hyperacusis also. Retraining is not simply an abstract learning exercise. In the subconscious part of the brain concerned with hearing, beyond the inner ear, but before the act of conscious perception of sound takes place, subconscious filters, networks of nerve cells (neuronal networks) are programmed to pick up signals on a "need-to-hear" basis.

Retraining therapy involves reprogramming or resetting these networks, which are selectively picking up "the sound life" in the auditory system. The presence of any continuous stimulus usually results in habituation whereby the individual responds less and less to the stimulus and in the final stage, the signal is no longer detected. This is achieved by retraining. TRT is based on strong neurophysiological evidence that any person can habituate to acoustic or acoustic-like sensations in their environment.

After knowing the basis of TRT - how is it used in hyperacusis individuals - ?

In these individuals TRT allows retraining lower parts of brain and inner ear to decrease their extreme sensitivity to moderate and loud sounds.

For the purpose, sound generators are used. These generators are housed in small ear level casing coupled with an open earmold. This does not interfere with patients hearing ability. These noise-generating instruments are worn in both the ears. They produce a gentle static-like sound (i.e.) a broadband noise (white noise). The volume control of devices are set so that noise is barely audible. This is in the initial part of the program. This level is maintained for six or more hours a day. When there are periods of uncomfortably loud environmental noise during the day, during such periods, the level of therapeutic noise should be turned up slightly to a level that begins to make the environmental noise less uncomfortable, but not nearly enough to mask it.

After a month, the level of noise should be raised progressively aiming to increase it gradually to higher and higher levels of tolerance - putting up with some discomfort (but not aggravating the tinnitus if present). This retraining period may vary from 6 months - 2 years. The advantage is that the use of sound generators makes them completely mobile and helps these individuals go about their work. There then comes a point where these individuals forget that they have their generators on. The disadvantage is its cost and the travel for regular follow-ups to a hearing professional.

Vernon at the Oregon hearing research center advices the use of low frequency sounds like pink noise than white noise and says that tolerance is developed better with low frequency sounds. This could be attributed to the reason that perhaps because white noise contains high frequencies at which these individuals dynamic range is inordinately small, thus creating irritation

and aggravation rather than leading to acclimatization. The environmental sounds to which we are exposed to normally contains little sound energy above about 4000Hz. So it is reasoned that use of "pink noise" (which lacks high frequencies) might improve the desensitization process by avoiding the high frequencies that are aversive to the hyperacusic patients.

To provide an appropriate listening experience that can be used by patients, special listening materials are available on cassette tapes or CD's. These materials can be listened to under earphones in order to provide maximum consistency and control of the sound level. Each time the individual wears the earphones and listens to pink noise, it is important that a proper, individualized listening level be established. The patient here is instructed to begin each listening session by increasing the loudness of the pink noise very gradually above threshold in order to determine the level at which the noise is just starting to be uncomfortable and then reducing the noise level only slightly. The patient has to then use that loudness level for the duration of the listening session, assuming that it remains comfortable.

The patient should be instructed that the goal is to gradually increase the listening level of the noise overtime, by raising each days listening level slightly relative to the preceeding day. Thus here the procedure takes advantage of increasing loudness tolerance as it starts to develop. Although it is told to the hyperacusic patient that it may take about 2 years to recover normal loudness tolerance, they may experience noticeable improvement within a few months.

Atleast a minimum of 2 hours is necessary to listen to pink noise. This can be divided into shorter intervals provided the listening level is established at the start of each listening period. Cessation of desensitization is recommended in cases where objectionable effects from listening to pink noise is seen.

The advantage to this approach is that it is inexpensive (price of CD or tape). The disadvantage is that the patient is confined to the noise source under earphones and is a self approach.

A study on experiences in the treatment of patients with tinnitus and or hyperacusis using the habituation method was done by Bartnik, G., Fabijanska, A., and Rogowski, M. (2001).

They designed a questionnaire to evaluate the effects of treatment. The questionnaire had to be answered before, during and after the treatment. The treatment was given for not more than a year. It was found by them that TRT appeared to be a successful therapy but it requires 18-24 months of treatment. They also said that further clinical studies on larger sample needs to be done to have a more precise assessment of the effectiveness of habituation.

Hyperacusis Instrument

The hyperacusis instrument developed these days works on the logic of extreme compression ratio and a low compression knee point (i.e.) low level sounds are amplified while the high intensity sounds are attenuated to quite an extent to avoid aversive condition in these individuals.

The electronic circuitry provides compression amplification that reduces or eliminates the amplification of higher level sounds. For low level sounds, the device provides carefully controlled amplification so that the wearer maintains a normal sound environment. The aim of these instruments is to maintain normal exposure to acceptable sound levels, thus avoiding overprotection of the ears.

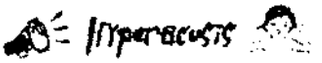
The similar logic has been used and various devices have been designed which include,

- Microtech has developed a hyperacusis instrument - a reverse hearing aid. The shell design uses a very tight fit and a soft ear canal and ensures that only sound entering the ear canal comes through the circuit.
- Jim Nunley, Jonathan lay and Gray son Silaski (cited in Vernon, 1998) are developing a special hearing aid (* 2000) with extensive compression so that sounds of any sort cannot rise above approximately 65dB SPL and very low intensity sounds are amplified up to a comfortable level that is adjustable. The prototype of * 2000 is being tested on hyperacusis patients and reports so for have been favourable.

  These instruments are available to any hearing aid dispenser or dispensing audiologist.
- Electronic loudness suppression devices - by Argosy Electronics Inc. is packaged in full - concha custom in-the-ear (1TE) housing. As

measured by Cabot Laboratories, the noise reduction rating for the earmold shell itself (with power off) was 28dB. An input compression limiter circuit with an extremely low compression threshold and a high compression ratio is used to keep maximum output SPL below loudness discomfort levels. There is variable release time to reduce the audible pumping action produced with a low threshold compression circuit. Benefits from the device have been reported. These devices are now not marketed by Argosy Electronics Inc.

The other management methods include relaxation techniques, biofeedback, and intake of some vitamin A tablets, anti-depressants etc but these lack scientific evidence.



HANDOUT FOR THE MANAGEMENT OF HYPERACUSIS FOR THE HEARING HEALTH CARE PROFESSIONALS

- What is hyperacusis ?

Hyperacusis is a condition where there is a collapsed tolerance to normal environmental sounds. It is widely acknowledged that hyperacusis involves a greatly reduced loudness tolerance for part or all of the frequency range, manifested in a small dynamic range.Sounds, or at least certain sounds, which would be easily tolerated by other persons are bothersome, annoying, uncomfortable or even painful to the hyperacusis patient.

- Is hyperacusis similar to recruitment ?

No. Hyperacusis and recruitment are two different conditions and one should not get confused between the conditions.

Hyperacusis is a collapsed tolerance to sound and generally have their hearing sensitivity within normal limits.

Recruitment is a condition of abnormal growth of loudness. It is a common feature of cochlear hearing loss.

- What are the causes leading to this condition ?

1. The origin of hyperacusis can be stated as to be in the central auditory system i.e. to say that the central auditory system gain or amplification for sound stimulation is excessively high.

2. It is also possible that there is a disruption in the efferent or descending pathway of the auditory system function. That is, without the usual amount of inhibition or modulation of cochlear activity by the efferent system,the brain is over stimulated by the acoustic sensory stimulation.

3. Another underlying condition for hyperacusis could be a reduction in a brain chemical called serotonin - This in someway influences a person's sensitivity or reaction to sensory stimuli.

4. Other causes could include

    - A sudden single burst of noise
    - A head injury
    - William's syndrome
    - Temperomandibular syndrome (TMJ)
    - Tay-Sach's disease
    - Meniere's disease
    - Bell's palsy
    - Lyme disease

- Airbag deployment
- Viral infections involving VIII nerve

• What is the audiological protocol to be used ?

A test battery should be used for audiological assessment

The battery should include

- Pure tone audiometry and oto-acoustic emissions to rule out sensory hearing impairment
- Some measure of word recognition
- Tympanometry to assess middle ear status
- Also hyperacusis should be evaluated carefully by both - direct frequency specific measurements of loudness discomfort.

  By a detailed assessment to detect any intolerance of specific environmental sounds.

• What can be offered to these patients as a hearing health care professional?

First and foremost step is to offer an understanding of the problem.

Hyperacusis is real for the patient and one can take a big therapeutic step forward by acknowledging the seriousness and validity of his / her problem.

One can take a thorough medical, audiologic and to a lesser extent, psychological history.Refer him / her to an audiologist for detailed assessment and management strategies.

• What is the role of an audiologist ?

The audiologist plays a very important role in counselling the patient and offering the available management strategies in an individualized fashion.

- Counselling plays a major role in the management of these clients.Reassuring the patient that the problem is not a serious one helps in relieving undue stress. An indepth discussion of the tests, anatomy and physiology of the hearing mechanism and how it relates to hyperacusis should be done. Educating the patient helps to relieve the fears and concerns present in the individual. Counselling needs to be part of the management program continuously.

- Ear protection can be another possibility. But care should be taken to use the ear protective devices only when necessary i.e. during exposure to high intensity sounds and for a short duration.

  In the initial stages, i.e. in cases where the onset is sudden, they can be recommended to use ear defenders, as they may be liable to further injury.

- Sound desensitization is yet another treatment avenue. Hyperacusis Habiruation therapy, which is based on Tinnitus Retraining Therapy, is recommended for these patients.

  For the purpose of sound desensitizarion white noise or pink noise generators can be used.

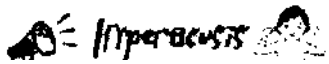
The patient should be counselled regarding the need to listen and get habituated to sound.

The client has to be trained regarding the use of noise generators.

The client plays a major role in this therapy once the program schedule has been made. The patient should also be told the time period that will be needed for the improvement to be noted.

- Lastly there are hearing instruments available for the condition, which works on the logic of extreme compression ratio and a low compression knee-point. These could be tried in appropriate cases.

  The audiologist can refer these patients to a psychiatrist if they have sleep disturbances and stress disorders related to hyperacusis



HANDOUT FOR THE MANAGEMENT OF HYPERACUSIS FOR THE LAYMAN

1. Imagine being at a movie where the sound track is turned to a very high volume. Actor's voices are screaming at you. After 5 minutes, you leave, holding your ears and cursing the theatre. Turning newspaper pages, running water, your child making noise - all are intolerable to your ears. A baby's cry or a screeching truck - The sound is too much - You ask, "what has happened to my ears ?".

If this is happening to you, you have a condition called "hyperacusis"

Hyperacusis is a condition when suddenly everyday sounds become unbearably or painfully loud.

2. What indications help me notice the problem ?

- You will feel that the loudness of particular sounds is too much, may cause pain. You may feel pressure on your eardrum.
- You may feel that sometimes you need to change your way of living because of your inability to bear loud noise.
- You may even find sounds of coughing, sneezing, snoring, clapping and laughing as loud.
- You may have to change your working environment if sounds become too loud for your ears.

- You may have difficulty even in sleeping
- You may sometimes find a change in your voice (talking softly, whisper or may be hoarse).
- You may not be able to use your household appliances.

3. Whom do I have to consult when I face the above ?

You can consult a hearing health care professional (ENT and Audiologist).He/She will take a detailed history regarding your problem. He/she will explain to you about your problem. For further management, the audiologist will help you.

4. What are the management strategies available for this condition ?

Not many solutions have come up for this problem. But the ones available may to some extent improve your condition. First and foremost

- Try not to move away from noise. Yes, it may be difficult at the initial stages, but slowly you will get over it.
- There are certain sound therapies available for this condition of yours.

In this method you will be required to listen to certain kind of noises for a period of time. Initially the noise will be at a level that is comfortable to you and slowly it will be raised. You have to listen to it everyday for a fixed duration over a period of time.

Once you are doing this, you are training your ears and your brain to accept noise. This may take sometime but do not lose hope.

- There are earplugs available. You can use them when you have to go to places likely to be excessively noisy such as rock clubs, vehicles etc. But, do not use them every time and continuously as you are going to worsen the condition of your ears. Once you get used to earplugs, you cannot live without it and this is not advisable.
- There are certain hearing instruments available which your audiologist can suggest. They cut down the noise and help you feel better and hence you can wear them on continuously.

5. Do I have a role to play ?

Yes, you definitely have a very important role to play. The audiologist will only guide you regarding what has to be done for your problem, but your acceptance of the problem and the way you are going to follow the guidelines will largely influence your improvement.

You can buy cassette / CD's of various noises that are available and use them at home regularly for the specified period of time. You also need to go for periodic follow-ups to your audiologist.

"Be patient" because eventhough this is a condition for which not many successful remedies are currently available, newer and better treatment avenues are being explored and may be made available sooner or later.

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