# Hearing impairments: causes, effects and rehabilitation



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# Hearing impairments: causes, effects and rehabilitation

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*David Baguley and Don McFerran*

## 1 Introduction

> There are two kinds of deafness. One is due to wax and is curable: the other is not due to wax and is not curable.
>
> Sir William Wilde (1815–1876)

It is almost 150 years since Sir William Wilde (Figure 1), one of the first surgeons to call himself an Otologist, and the father of the celebrated writer, Oscar, considered the causes of deafness in this pessimistic way. The present situation is very different, and in this chapter the causes of hearing impairments and the many strategies that can be utilized to rehabilitate hearing-impaired individuals will be considered. Hearing impairments are normally divided into groups based on the anatomical site of the problem. Thus hearing losses can be subdivided into conductive (outer ear or middle ear), cochlear (inner ear), **retrocochlear** (auditory nerve) and central (brain) causes. The standard tests of hearing easily distinguish conductive losses from the other types but are less good at distinguishing cochlear, retrocochlear and central auditory problems from each other. The latter three causes are therefore in practice often amalgamated and the classification is simplified to conductive and sensorineural.

**Figure 1** Sir William Wilde (1815–1876).

## 2 Conductive deafness

### 2.1 Causes

The **conductive pathway** comprises the pinna or external ear, the external auditory meatus or ear canal, the tympanic membrane or eardrum and the middle ear. The middle ear is the small cavity between the eardrum and inner ear, which contains the ossicles. The ossicles are the three tiny bones of hearing, the malleus, incus and stapes or hammer, anvil and stirrup. Disease of any part of this pathway can result in hearing impairment.

#### *Pinna*

The pinna can be congenitally deficient or can be damaged by trauma or tumour. Although the pinna has a small effect in funnelling sound and contributing to sound localization it is not of major importance in humans. Some other mammals, such as horses and cats, have more highly developed external ears and have a much enhanced ability to localize sound accurately. Disease of the human pinna can cause marked cosmetic deformity but generally does not cause significant deafness.

#### *External auditory meatus*

The external auditory meatus can become blocked due to wax or a foreign body, or it can become swollen due to infection or inflammation. This impedes the passage of sound to the ear drum. It is easy to mimic this hearing impairment by simply putting your fingers in your ears!

### *Tympanic membrane*

The tympanic membrane can be damaged by trauma, such as a slap to the side of the head, or by fluid resulting from infection in the middle ear (called **acute otitis media**) bursting its way out through the membrane (effusion). These factors can cause holes or perforations of the eardrum, which reduce the amount of sound energy that is captured by the ear thereby causing hearing loss (Figure 2). The eardrum can become scarred as a result of repeated infection. This scarring, referred to as **tympanosclerosis**, increases the stiffness of the membrane, reducing its efficiency as a sound transmitter.

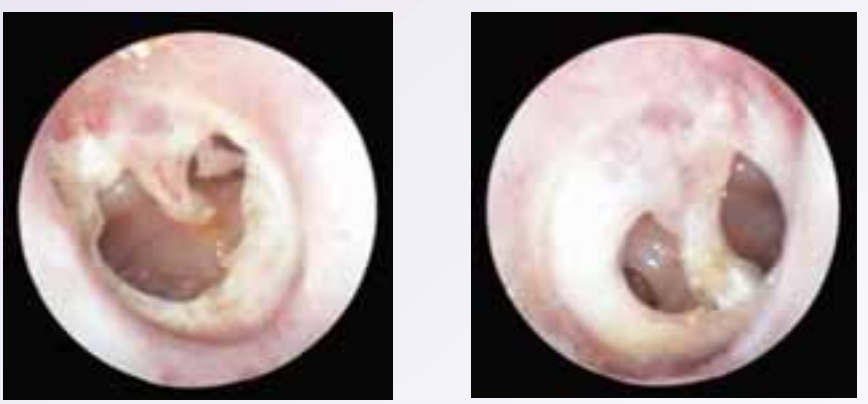


**Figure 2** Subtotal perforation of tympanic membrane in left and right ears.

### *Middle ear*

The middle ear cavity normally contains air which reaches this cavity via a small tube called the Eustachian tube that passes from the back of the nose up to the ear. If this tube malfunctions, air cannot reach the middle ear cavity and the ears feel blocked – most people have experienced a temporary blocked feeling of the ears when they have had a bad head cold. If the tube malfunctions for a longer period, the middle ear cavity gradually fills up with fluid that is produced by the lining of the cavity itself. This fluid may be thin and watery or it may be the thick, viscid (sticky) fluid that gives the condition the colloquial name ‘glue ear’. The more correct medical terms are **otitis media with effusion** or **secretory otitis media**. Although otitis media with effusion can occur at any age it is most prevalent in children partly due to immaturity of the Eustachian tube and partly due to the physiological enlargement of adenoid tissue in this age group. The adenoids are swellings of lymphoid tissue at the back of the nose that help the body to respond to infections. Unfortunately the adenoids are very close to the openings of the Eustachian tubes and adenoidal swelling can result in tubal blockage. Otitis media with effusion is by far the most common cause of conductive hearing loss in the UK.

### *Ossicles*

The ossicles can be damaged by infections; acute otitis media is a common condition in childhood that, in addition to causing perforations, can result in scar tissue developing round the ossicles. Persistent negative pressure in the middle ear can suck part of the eardrum inwards. The inside-out portion of the eardrum traps layers of dead skin that can then become infected. This small bag of chronic inflammation is called a **cholesteatoma**, and the infectious material slowly,

inexorably, eats away the normal structures in the middle ear. The middle of the three ossicles, the incus or anvil, is especially prone to damage in this process. This uncommon disease tends to occur in later childhood or adulthood. Trauma can also damage the ossicles; a severe head injury can dislocate the delicate joints between the tiny bones. In the condition of **otosclerosis** new bone forms around the footplate of the stapes, fusing the bone rigidly with the bone of the inner ear. This reduces the ear's ability to conduct sound from the ossicular chain to the transduction mechanism of the cochlea.

## 2.2 Effects

Even with no ear canal, eardrum or ossicles, sound can still reach the inner ear, transmitted through the bones of the skull directly to the cochlea. The maximum effect of a conductive lesion, therefore, is to remove the contribution of the conductive pathway to hearing. An isolated conductive hearing loss has a maximum extent of 40–50 dB.

In addition to measuring the individual's **hearing thresholds** (the quietest sounds they can hear in a sound-proofed environment) it is also necessary to consider **auditory discrimination** while assessing hearing impairment. This is the ability to listen to a sound in a noisy environment and to be able to concentrate on that sound while ignoring the background noise. An individual with reduced hearing sensitivity but normal auditory discrimination will be helped simply by making the sound louder. In contrast, an individual with poor auditory discrimination will find that simple amplification will turn a quiet acoustic jumble into a louder acoustic jumble. The clarity may not be improved. Isolated conductive hearing losses generally have normal auditory discrimination.

## 2.3 Rehabilitation

As they have normal auditory discrimination, individuals with conductive hearing impairments generally do well with hearing aids, but herein lies a great irony, as this is the same group of people that are most likely to benefit from surgery. An offending foreign body or wax can be removed, a perforated tympanic membrane can be repaired with a graft taken from one of the patient's own muscle tendons, and a dislocated ossicular chain can be reset using microsurgical techniques. Cholesteatomas can be excised and the damage they have caused can in many cases be repaired. In cases of otosclerosis the diseased stapes can be replaced with a minute prosthetic ossicle, such stapedectomy surgery often returning hearing sensitivity to normal or near normal levels.

The situation regarding cases of otitis media with effusion has been the cause of much debate and controversy in recent years. The persistence of this condition in a child can undoubtedly impair that child's development, both in educational terms and also in social skills. Surgery has been developed to alleviate the potential handicap. The surgery involves making a tiny incision in the tympanic membrane, removing any fluid in the middle ear and inserting a tiny plastic or metal tube. This ventilation tube or **grommet** (Figure 3) allows air to get from the atmosphere, through a hole in the grommet into the middle ear cavity. This equalizes the pressure on either side of the eardrum, preventing build up of fluid in the middle ear. The tympanic membrane grows throughout life and this normal growth eventually results in the grommet being extruded from the eardrum. This process takes approximately one year and in the majority of cases, by the time extrusion is complete, the patient's Eustachian tube function has returned to normal and the otitis media with effusion does not recur.

**Figure 3** A T tube and Shah grommet.

Initial results were good and the operation became very popular. However, more detailed surveys showed that the otitis media with effusion was short lived in many cases and the insertion of grommets was not without its own problems; scarring of the eardrums often followed grommet insertion and in some cases the degree of hearing improvement was disappointing. In addition, the operation developed a political dimension: grommet insertion became the most frequently performed surgical procedure in the UK with associated financial implications. Healthcare purchasers therefore sought to reduce the number of grommet operations. A sensible middle ground approach has now been adopted whereby most individuals with otitis media with effusion undergo a period of 'watchful waiting' (careful monitoring of a child's hearing over time) prior to consideration of surgery. In some Otology units hearing aids are provided for mild cases, surgery being reserved for the more seriously affected.

# 3 Cochlear hearing loss

## 3.1 Causes

The delicate micro-mechanical structures of the cochlea have been described previously (Chapter 2, *The transformation of sound stimuli into electrical signals*) and it will come as no surprise that the cochlea is vulnerable to trauma, infection and degenerative processes, leading to reduction of hearing sensitivity. Noise at excessive levels, even for short periods of time, affects first outer and then inner hair cell function. A decrease in cochlear sensitivity with age is almost universal, the consequent hearing loss being entitled **presbyacusis**. **Menière's disease** is a disorder of the inner ear characterized by episodes of hearing loss, tinnitus and vertigo. The ear often feels blocked prior to these attacks. Between acute episodes the patient may return to normality though as the disease progresses the hearing loss may become permanent. This distressing condition is thought to be due to a build up of endolymph within the inner ear that distorts the delicate membranes of the cochlea and vestibular apparatus, thereby affecting both hearing and balance. In some rare cases the body's immune system may malfunction, attacking normal tissues such as the components of the inner ear causing a sudden or progressive hearing loss. The cochlea is also vulnerable to a number of cochlear-specific viruses. Congenital cochlear impairment will be considered as a separate entity.

## 3.2 Effects

The extent of cochlear hearing loss can be mild, moderate, severe or profound, and can have almost any conceivable frequency configuration, though most often the extent of loss is greater in the high frequencies. With cochlear deafness due to ageing or noise exposure the loss is usually bilateral and reasonably symmetrical. Some other forms of cochlear loss, including deafness due to Menière's disease or trauma can result in unilateral symptoms. Auditory discrimination tends to be more affected in this type of loss than in conductive losses.

## 3.3 Rehabilitation

In Menière's disease, specific but controversial treatments are used to prevent or ameliorate attacks. Drugs are used to try and improve cochlear blood flow and also to control the vertigo. Surgery is undertaken in some patients to try and reduce the build up of endolymph. Other patients undergo operations to sever the balance nerves and prevent the disabling vertigo.

However for the majority of patients with cochlear hearing loss there is no operative or drug treatment. The prevalence of cochlear hearing loss in the UK has been investigated, and it appears that about one in ten adults would benefit from hearing aids. Unlike conductive hearing impairment the effect of cochlear damage is not only to reduce hearing sensitivity, but also to reduce the ability to make fine discrimination in frequency and temporal domains. In addition, tolerance to loud sound may be reduced. The consequence for an individual is that sound may be distorted, even when amplified. Thus the benefits of hearing aids in such situations may be limited unless they are carefully prescribed to account for an individual's specific needs in this regard. There are some early indications that hearing aids which utilize digital technology rather than analogue may be better able to meet the requirements of a cochlear hearing-impaired person, though this is as yet unproven. What is generally accepted is the need to match the prescribed hearing aid sound output with the needs of the patient (Figure 4).

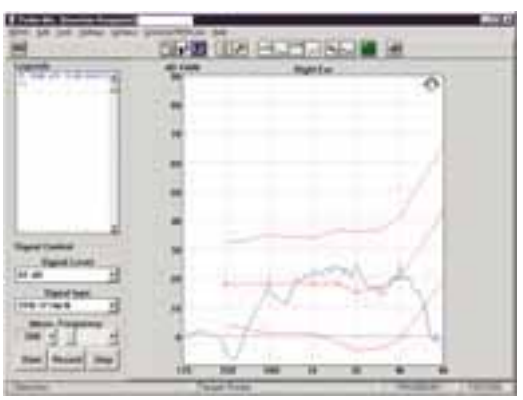

**Figure 4** Computerized hearing aid fitting screen, demonstrating hearing aid performance measured in the ear canal of the patient. The blue line represents the hearing aid response, the central red line depicts the ideal response for this patient.

Whilst technological solutions to cochlear hearing impairments are advancing, it should be noted that these do not address the whole problem created by the hearing impairment. Adult individuals who acquire a cochlear hearing loss may experience feelings of isolation and loss of confidence, and a small but significant proportion may go on to experience psychological problems as a result of their hearing impairment. The application of technology to address this in an holistic context appears to be the most effective method of intervention, thus treating people as people rather than dysfunctional hair cells! It is noteworthy however that the opportunity to perform technological and holistic rehabilitative interventions is largely limited to the developed world. The possibilities of cheap and appropriate interventions, such as solar-powered hearing aids and campaigns for the prevention of hearing loss are being investigated for the benefit of the large number of hearing-impaired people in the Third World.

## 3.4 Congenital cochlear impairment

In the developed world approximately 1 in 1000 live births is a child with a bilateral severe to profound cochlear hearing loss. In approximately two-thirds of these cases the cause of the problem is genetic. In the other third, factors such as intrauterine infection, hypoxia or neonatal jaundice are responsible. Without early identification and treatment such children have almost insurmountable difficulties in the development of speech, and sign language may become vital for communication. Early identification is possible with tests undertaken in the neonatal period, though care has to be taken to allay parental anxiety. Initial treatment of such children is to provide suitable hearing aids, intensive support for both child and parents, and to

correct any concomitant conductive hearing loss such as otitis media with effusion. In some cases however, the hearing loss is beyond the reach of even the most powerful and sophisticated hearing aids.

The idea that it is possible to induce sound perception by electrical stimulation dates as far back as Volta who in 1800 undertook electrical stimulation of his own ear after filling the ear canal with saline solution, the electrical charge giving rise to an extremely loud bang – Volta appears not to have repeated the experiment! In the 1970s the possibilities of using electrical stimulation to give the profoundly deaf some experience of sound formed the basis of **cochlear implantation**. Those early devices utilized a single electrode placed in the round window to stimulate the cochlea. Nowadays a multi-channel electrode is usually placed within the cochlea, and complex speech-processing algorithms are used to stimulate the auditory nerve, giving as rich and natural a perception of sound as possible (Figures 5 and 6).

**Figure 5** The Clarion® range of external cochlear implant equipment made by the Advanced Bionics Corporation. The sound signal is converted to an electrical signal by a microphone attached to a microprocessor worn either on the belt (top) or behind the ear (bottom, shown with attached headpiece). After processing, the signal is sent to a headpiece, which sends the sound information through the skin to the implant using a radio signal.

**Figure 6** Schematic diagram of the insertion of a cochlear implant electrode into the cochlea.

Initially application of cochlear implants was in deafened adults, many of whom had lost their hearing as the result of meningitis, but cochlear implant surgery is now routinely undertaken in children born with a profound hearing loss.

Patients with a cochlear implant find that at the least the device augments their abilities to lip-read and increases awareness of environmental sound, whilst at best they are able to converse on the telephone and to enjoy music. The reasons for such variability in performance are not fully understood but include the length of time a person has been deaf, the disorder causing that deafness and the number of surviving cells in the auditory nerve. One important factor for congenitally deaf children in the consideration of cochlear implants is the age of the child. It is widely accepted that after the age of approximately eight years the neural plasiticity needed for the auditory system to utilize stimulation from a cochlear implant may not be sufficient to justify this intervention. Cochlear implants do have their opponents; some deaf adults think that cochlear implants are an act of violence upon deaf children, whose natural language is said to be sign language and whose natural home is in the deaf world. Despite this however, developments in the field continue, and it is expected that binaural cochlear implantation may soon become commonplace.

# 4 Retrocochlear hearing loss

## 4.1 Causes

The cochlear and vestibular nerves run side by side within the internal auditory meatus (the bony canal that passes through the temporal bone from the inner ear structures towards the brainstem). A region of cellular instability on the vestibular nerve can give rise to a benign tumour, derived from the Schwann cells that surround and insulate nerve fibres. These tumours are correctly known as **vestibular schwannoma** but are more often referred to as **acoustic neuromas**.

## 4.2 Effects

Vestibular schwannoma usually cause unilateral hearing loss and tinnitus (Section 6). The hearing loss typically results in poor auditory discrimination. Left untreated they can grow to fill the internal auditory meatus, and thence grow out towards the brainstem, where after some years the compressive effects of the tumour compromise the functions vital for life. Vestibular schwannoma are diagnosed in 1 in 50 000 adults per year in the UK, and such diagnosis has been greatly facilitated by magnetic resonance imaging (Figure 7 overleaf).

## 4.3 Treatment and rehabilitation

At present the usual treatment of this condition is the surgical removal of the tumour, undertaken by a team including both otologists and neurosurgeons, though technologies involving the irradiation of the tumour are being investigated. The surgical procedures commonly employed for this condition cause a unilateral total and permanent hearing loss in the operated ear.

## 4.4 Other retrocochlear conditions

The move towards neonatal diagnosis of hearing impairment has uncovered another situation of interest. In the 1970s biophysicist David Kemp working in London discovered that normal ears emit a small amount of sound in response to sound

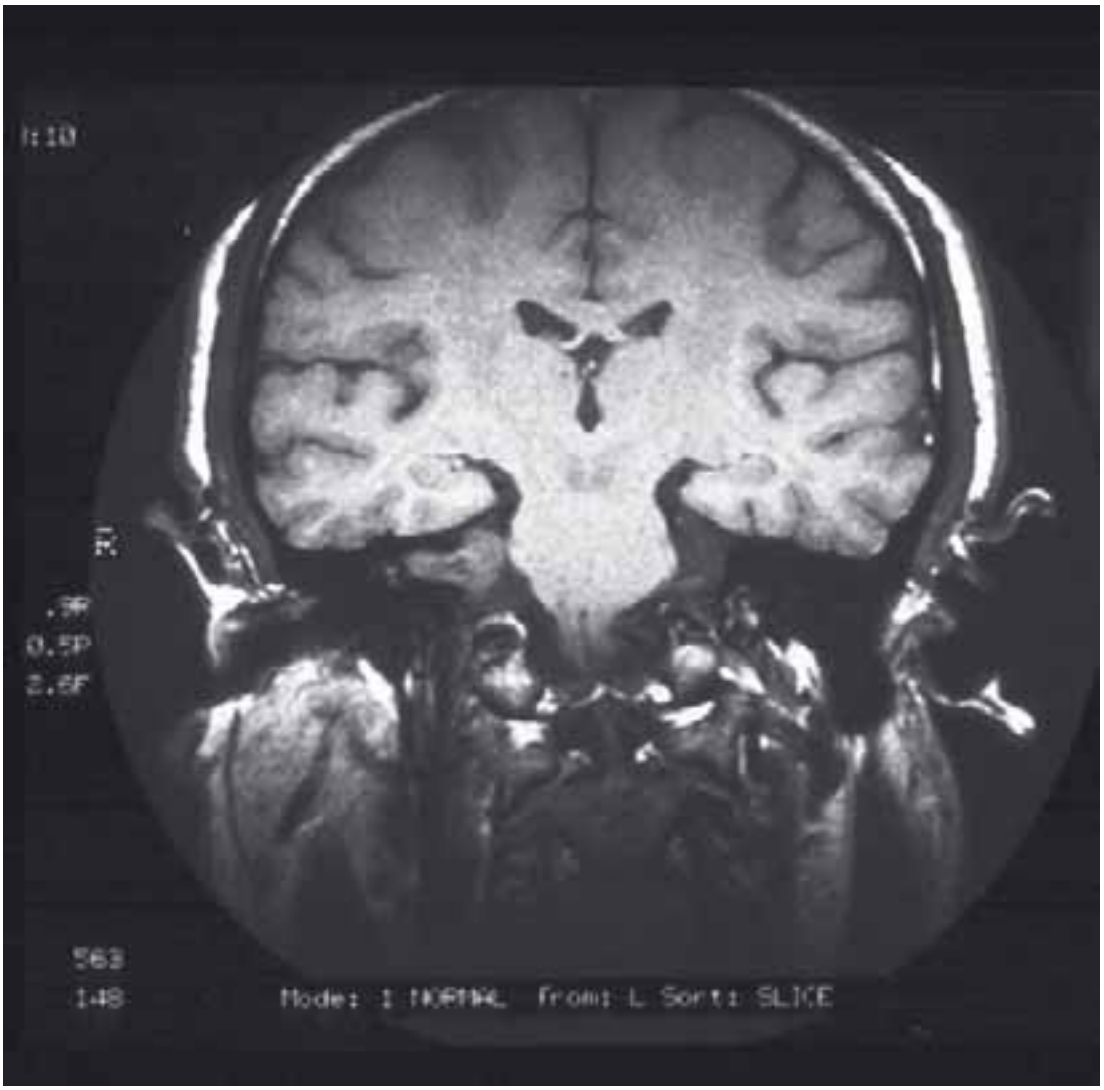


**Figure 7** MRI scan demonstrating right vestibular schwannoma.

input. These **otoacoustic emissions** are used as the basis of a hearing test for neonates (newborn infants). A small number of babies have normal otoacoustic emissions suggesting normal cochlear function, but when the activity of the retrocochlear neural pathway is tested using auditory brainstem responses the results are indicative of significant dysfunction. Further investigation has shown that the auditory nerve fibres seem to be unable to respond to sound in a synchronous fashion. This condition has been entitled auditory neuropathy or auditory desynchrony, and is a compelling argument for using a battery of tests for the diagnosis of hearing impairment in young children rather than relying upon one test alone. Similar patterns of auditory nerve dysfunction may be demonstrated in 20% of adult patients with multiple sclerosis, though it is rare for such patients to complain of hearing difficulties.

# 5 Central auditory processing disorders

## 5.1 Causes and effects

One interesting group of patients that are referred to audiologists and otologists believe themselves to have a hearing problem although standard hearing tests are normal. More detailed psychoacoustic tests show that their ability to discriminate sound in noise is markedly worse than their peers. In the UK these symptoms were carefully considered in the 1980s and the condition named **Obscure Auditory Dysfunction** (**OAD**). In the USA the name **Central Auditory Processing Disorder** (**CAPD**) has been used, and was coined after the specific investigation of children who had normal hearing acuity on pure tone and otoacoustic emission testing but who had quantifiable and significant difficulties in noisy environments – such as found in classrooms. CAPD is gradually becoming the more accepted terminology. Specific mechanisms involved in this situation have yet to be identified, and indeed there are likely to be a variety of different patho-physiologies involved rather than one alone.

## 5.2 Rehabilitation

Management of CAPD involves a variety of measures. Increasing the strength of the signal that the individual is striving to hear relative to the competing background noise is the first avenue to investigate. In a classroom this can be accomplished by simple measures such as moving the child closer to the teacher. Careful use of hearing aids or FM radio aids can also help. For adult sufferers it is often more difficult to manipulate the acoustic environment and in these cases counselling and the teaching of specific hearing tactics are the most common treatment options.

# 6 Tinnitus

Psychologist Dennis McFadden in 1982 defined **tinnitus** as 'the conscious expression of a sound that originates in an involuntary manner in the head of its owner, or may appear to him to do so'. It has been determined that approximately one-third of the UK adult population experience some short-lived (< 5 minutes) spontaneous tinnitus experiences and that in 10% tinnitus experience is persistent. In 1% of the UK population tinnitus has a severe effect upon quality of life, making this an auditory dysfunction worthy of scientific consideration and of effective therapy. Unfortunately, despite considerable scientific endeavour (there being nearly 1700 peer review scientific papers considering tinnitus in the last decade) effective treatments remain elusive. Drug therapy has not been effective, although antidepressant medication can help to alleviate any depression associated with tinnitus. Surgery is not usually recommended, although some conditions that may require surgery (such as vestibular schwannoma and Menière's disease; see above) contain tinnitus within their symptom profile. Treatment strategies rely upon counselling, and the reduction of sympathetic autonomic nervous system (agitation) and limbic system (emotion) responses to tinnitus. A useful model of tinnitus was proposed by neuroscientist Pawel Jastreboff in 1990 (Figure 8 overleaf), pointing out that whilst the generator site of tinnitus may be in the auditory periphery (though not necessarily the cochlea) the mechanisms of persistence and reaction were central and also involve the sympathetic and the limbic system (specifically the amygdalae, structures in the brain that play a critical role in the formation and modulation of emotionally influenced memory).

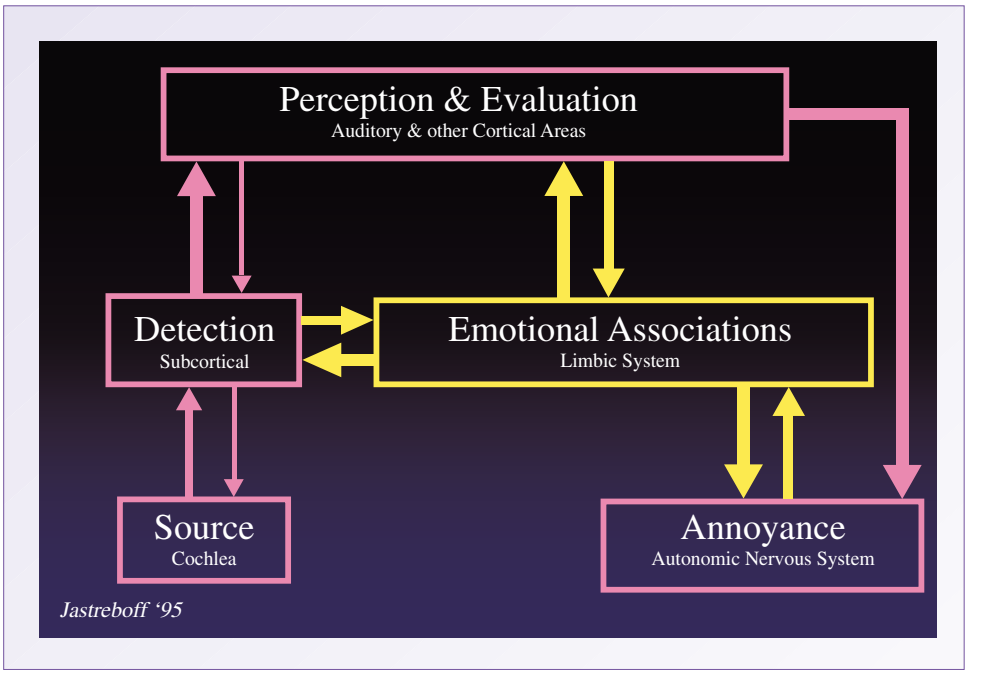


**Figure 8** Jastreboff neurophysiological model of tinnitus. The model postulates that perception of tinnitus becomes associated with negative emotions, thereby involving the limbic system, which in turn activates the autonomic nervous system, resulting in annoyance.

Whilst this model has been criticized for offering little new scientific insight, the clinical impact has been high, with a treatment strategy named 'Tinnitus Retraining Therapy' based upon the model. This approaches the treatment of tinnitus from several different angles; the condition is thoroughly explained, the patient is counselled, relaxation techniques may be taught and sound therapy is used in some cases (Figures 9 and 10).

As yet there are no good properly controlled scientific trials of this therapy but there is considerable ongoing research in the field. There may be some controversy over the methods of tinnitus therapy, but there is a consensus about the objective, that patients should habituate to the signal within their auditory system. One is unlikely

**Figure 9** Environmental sound generator used in sound therapy for tinnitus.

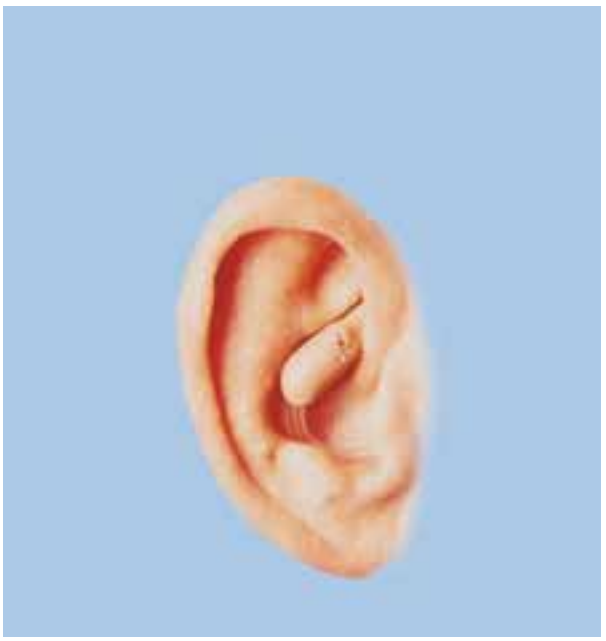

**Figure 10** Ear-level wide-band noise generator used in tinnitus therapy.

to do so if anxious or upset by tinnitus, or concerned about the potential seriousness of the sound, and so careful, authoritative reassurance might well be helpful, and indeed has long been utilized by some audiologists and otologists.

Some recent scientific insights into tinnitus may well augur more hope for the future. Whilst tinnitus may well emerge as a consequence of changes in cochlear function, it has been suggested that such changes result in reorganization of the auditory cortex, specifically in regard to the dimensions of the cortex concerned with the perception of particular frequencies close to an area of hearing loss. An analogy is drawn with the situation of losing a finger, wherein the area of the somatosensory cortex concerned with that finger is greatly reduced in activity immediately following that amputation. Some weeks later activity has returned to usual levels, but investigations have shown that this later activity is associated with neighbouring fingers which are thus over-represented in terms of cortical activity. This phenomenon has been associated with the development of phantom pain and the suggestion is that tinnitus represents phantom auditory perception in some cases. Investigation of such hypotheses continues apace, and the hope is that some more effective therapy for people who develop tinnitus may result.

## 7 Summary

The human auditory system is complex and dysfunction at any level may result in hearing impairment. The simple reduction in sensitivity in conductive hearing loss may be remediated by amplification, but the more complex dysfunction associated with cochlear hearing loss requires careful consideration. Auditory dysfunction beyond the cochlea is also possible, and may result in particular auditory phenomena. Tinnitus is common in the population in general and appears not to bother most of these individuals, but as there is a significant number of people whose lives are significantly affected, this condition therefore warrants careful consideration and the development of effective therapy.

## Further reading

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