SUDDEN DEAFNESS: A REVIEW OF LITERATURE (1989-1998)

Register No.M9806

Independent Project submitted as part fulfilment for the FIRST Year M.Sc, (Speech and Hearing), submitted to the University of Mysore, Mysore.

ALL INDIA INSTITUTE OF SPEECH AND HEARING: MYSORE 570006

MAY 1999

Dedicated to

Amma and Appa

With love and

gratitude

CERTIFICATE

This is to certify that this Independent Project entitled : **SUDDEN DEAFNESS : A REVIEW OF LITERATURE (1989- 1998)** is the bonafide work in part fulfilment for the degree of Master of science (Speech and Hearing) of the student with Register No.M9806.

Mysore
May, 1999

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CERTIFICATE

This is to certify that this Independent **Project** entitled : **SUDDEN DEAFNESS : A REVIEW OF LITERATURE (1989-1998)** has been prepared tinder my supervision and guidance.

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DECLARATION

This Independent Project entitled : **SUDDEN DEAFNESS : A REVIEW OF LITERATURE (1989-1998)** is the result of my own study under the guidance of Dr.Raj alakshmi K, Lecturer in Audiology, All India Institute of Speech and Hearing, Mysore and has not been submitted earlier at any University for any other diploma or degree.

Mysore
May, 1999

Reg. No.M9806

ACKNOWLEDGEMENT

My deepest gratitude, reverences and heartfelt thanks to my guide Dr. Rajalakshmi K. Lecturer in Audiology, All India Institute of Speech and Hearing, Mysore, for guiding me throughout this project work and being a strong moral support to me through all the stages of this project work.

I sincerely thank Dr. (Miss) S.Nikam, Director, All India Institute of Speech and Hearing, Mysore for granting me permission to take up this project.

My dearest Appa and Amma - I consider myself blessed to have parents like you. Your prayers and encouragement have always been contributing in urging me to move ahead and handle all the obstacles coming on the way with courage. I owe my life to you.

Dearest T.V.M.ammai : It was your encouragement and moral support on the basis of which I had come here for my further education. Words fail to thank you forall the support you have provided during those 'critical' moments. We (Appa, Amma, Sharath, Achyuth, Sreekanth and I) can never repay you for all that you have done to our family. I am so glad I have you as my 'Granny' !

Dearest Sharath : A "wonderful loving, caring brother" is what I have seen in you. Though miles apart, your constant encouragement has been a strong support to me. Are words sufficient to say "Thanks"? for being such a wonderful brother, whom I know I can count upon in any stage of my life.

Dearest Achyuth and Sreekanth - you both younger ones form a cute little pair whom any one would love to have as their brothers. I am so glad you are mine !

Dearest N.K.M. I thank the Almighty for letting me know sucha wonderful person like you. You are the invaluable "treasure" of my life and I am glad I have found you. Though miles away, oceans apart, you never failed in providing me all the support and encouragement I needed. Can I ever thank you enough for all that you have added to my life?

Kusum : It's you who have let me know the meaning of the word "FRIEND". Your lovely letters have always been a constant source of joy and encouragement. Eagerly awaiting for those golden moments of friendship to be back again.

Milind : You have been constant source of support and encouragement throughout my stay here. "Thanks" for being such a wonderful and caring senior and friend.

Dearest Prarthana and Uma: A special" thanks" to you both for being wonderful roommates who always made me feel "at home".

A special note of thanks to all my dear classmates for all the "golden moments" shared together.

I also thank Rajlakshmi akka for her quick and efficient typing work and enabling me to finish the project work in time.

I thank all the people who directly or indirectly have helped me in finishing this project!

Last but not the least. I thank the" ALMIGHTY" for bestowing me with a wonderful life and for always being with me in my joys and sorrows.

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INTRODUCTION

When people refer to the ear they generally think of those structures that protrude off the side of their heads and serve to support eyeglasses and ear-rings. However, the ear is actually a much more complex structure that is divided into external, middle and inner components.(cited in http:www.ent.utmem.edu/restibulo coch/ shl.htmb).

The external or outer ear consists of that portion of the ear that protrudes off the side of the head together with a long tunnel, known as the external ear canal, that leads to the eardrum. The external ear serves to conduct sound to the middle ear. Wax is produced in the ear canal to protect the skin of the canal from infection and water damage. *.(cited in http://www.ent.utmem.edu/restibulococh/shb.htm)*

The middle ear begins at the eardrum, which is a thin sheet like structure that completely seals the middle ear from the external ear. Air passes to the middle ear from the back of the nose through a long tube known as the eustachian tube *.(cited in http://www.ent.utmem.edu/ restibulococh/shb.htm).*

The inner ear is the critical organ responsible for hearing. While the external and middle ear amplify the sound, it is the inner ear that converts sounds into electrical energy that can be transmitted to, and processed by the brain. Essentially, the middle ear transmits two forms of information to the brain concerning sound. The first piece of information concerns the intensity or loudness of the sound. The second piece of information concerns the frequency or pitch of the sound. It is the information concerning the unique pitches or frequencies of different sounds that enables the brain to understand complex sounds such as

words and music. Loss of ability to discriminate different pitches of sounds results in a decreased ability to understand complex sounds such as words.

From the inner ear sound passes to the brain via a nerve known as the "auditory nerve".

Thus, we may conclude that the ear is the primary sense organ of hearing and any effect on the external, middle or inner ear may lead to either a conductive, mixed or sensori-neural hearing loss.

The term "hearing loss" is used whenever a specific reference is being made to a hearing-impairment which is of particular intensity magnitude such as a 40 dB hearing loss. The term "deaf' refers to persons in whom the sense of hearing is non-functional when used alone, with or without amplification, for the ordinary purposes of life. Such an individual may

a) have been born either totally deaf or sufficiently deaf to prevent the establishment of speech and natural language.
b) have become deaf in childhood before language and speech were completely established (prelingual) or
c) have become deaf after acquiring speech and language skills (post lingual), thus significantly impairing communication skills (Nicolosi, Harryman and Krescheck, 1978).

Hearing loss resulting from a dysfunction of the external ear leads to a conductive hearing loss, the causes of which may be (i) congenital disorders such as pure canal atresia (absence of external

auditory canal), which may be associated with malformation of the auricle such as microtia (small auricle) anotia (absence of auricle), etc. (ii) acquired causes such as infections of external ear canal (otitis externa), foreign body obstruction of the ear canal, growths of the bony external canal such as osteomas etc.

Dysfunction of either the external or middle ear may lead to a mixed hearing loss, the cause of which can either be congenital such as fused middle ear ossicles, absence of stapedius tendon, aberrant courses of the facial nerve or acquired infections such as in chronic otitis media, aero otitis media etc.

Disorders of the inner ear may lead to a loss of the sensorineural type, the causes being congenital such as following maternal rubella in the first and second trimesters of pregnancy, drug ingestion by the mother (eg. thalidomide, quinine) etc. Acquired causes leading to sensorineural hearing loss may either be due to Meniere's disease, due to presbycusis (degenerative changes of aging), or induced by drugs (such as streptomycin, neomycin, etc) or viral infections (eg. mumps).

Hearing loss may also have a sudden onset in which case it is known as a sudden hearing loss. This may result either from a dysfunction of the external or middle ears (leading to a sudden conductive hearing loss) or from a dysfunction of the inner ear or the auditory nerve (leading to a sudden sensorineural hearing loss).

(The most common causes leading to a sudden conductive hearing loss include wax impacted in the ear canal, infection of the external or

middle ear, fluid accumulation within the middle ear. Fluid accumulation may result from sudden changes in air pressure, such as those experienced on an air flight. A physical examination will confirm the presence of abnormalities within the external or middle ear and treatment can be administered to address these deviations. This may include removal of wax, or treatment with antibiotic or decongestants.

(The vast majority of cases reported with sudden hearing loss are primarily of the sensorineural type. Although usually having a gradual onset with slow progression, sensorineural hearing loss may also have a sudden onset. The onset may be instantaneous or the loss may progress over hours or days. A roaring tinnitus may accompany the deafness, and vertigo is not uncommon. Sudden sensorineural hearing loss may be due to a number of known causes of hearing loss including bacterial labyrinthitis, meningitis, encephalitis, multiple sclerosis, syphilis etc. Therefore, a thorough search for potentially treatable causes of sudden sensorineural hearing loss must be undertaken with an exhaustive history, thorough physical examination, audiological evaluation and carefully selected laboratory and radiographic studies)

The existence of rapidly progressive forms of sensorineural hearing loss (sudden hearing loss and rapidly progressive sensorineural hearing loss) constitutes a challenge in terms of the etiopathogenic diagnosis of this group of disorders. It is difficult to pinpoint the temporal limits that distinguish between these different clinically reversible forms, but it is widely accepted that sudden deafness develops in less than 72 hours, is generally unilateral and resolves spontaneously in one-third of the patients.

Sudden deafness has been investigated thoroughly in recent years. It emerges suddenly and yet some of the cases are spontaneously cured, despite the fact that it is a type of sensorineural hearing loss. Most cases of sudden deafness remain idiopathic and the majority are unilateral. Sudden hearing loss is a frustrating and frightening condition especially if the hearing loss is bilateral.

Sudden deafness is involved in about 1% of all cases of sensorineural hearing loss (JafFe, 1973) and more than 100 causes have been found for sudden deafness. Thrombosis, Vasospasm and viral infection have usually been proposed as causative factors. However, the etiology of sudden deafness remains incompletely understood.

Friedrich (1985) found a high incidence of cardiovascular risk factors and Belal (1980) found cochlear sclerosis in his cochlear tomography studies, about one third of the patients suffer from sudden hearing loss of viral origin (Jaffe, 1973). Many theories have been put forward regarding the cause and pathogenesis of this enigmatic symptom.

Disturbance of the blood circulation of the inner ear has long been one of the main causal hypotheses. The internal auditory artery is an end artery, making the inner ear very susceptible to damage to this artery. As early as in 1944, de Kleyn postulated a central vascular lesion in many of the cases that he reported. Some authors have attributed these symptoms to a deficient homeostatic system, often in conjunction with general risk factors for vascular disease and arteriosclerosis. Other researchers have found a relationship between high blood viscosity and

sensorineural hearing loss in general; it has recently been shown that sudden deafness is connected with high blood viscosity. Others again have postulated a viral cause discussing a possible virus induced detrimental effect on the rheologic properties of the blood. In short, there are many factors that could lead to thrombosis formation or emboli in the cochlear vessels.

Every patient suffering from sudden hearing loss must be thoroughly evaluated in order to treat any underlying disorders that may be contributing to this disorder. This assessment begins with a comprehensive history which should outline the complaint (speed of onset, duration of the hearing loss and associated symptoms such as vertigo, tinnitus, auralfullness, headaches, vision changes). Special attention should be diverted towards the otolaryngologic and neurologic portions of the examination. The history may alert the ear, nose and throat doctor to the situation which may have caused the loss, such as upper respiratory infection, air travel, history of smoking, taking of birth control pills, or other ototoxic medications and existing medical disorders. .*(cited in http://www.hearing center.com'info-files/sudloss.htnl).*

The ear, nose and throat examination will show any abnormalities such as excessive wax in the outer ear canal infection or fluid in the middle ear, or perforation of the ear drum.
.*(cited in http://www.hearing center.com/info-files/sudloss.htmb)*

Blood tests included in the assessment help to check for elevated levels of triglycerides and/or cholestrol, syphilis, infection, diabetes, thyroid and kidney disorders, inner ear infections and auto-immune disease..*(cited in http://www.hearing center.com/info-files/sudloss.html)*

An audiological evaluation will determine the degree type and location of the hearing loss, whether it is a loss of the middle ear, inner ear or auditory nerve. In addition, testing will be completed to determine the ability to discriminate speech in quiet as well as in noise. *.(cited in http://www.hearing center.com/info-files/sudden loss.html)*

Immittance testing may also be completed to assess the physical status of the eardrum and middle ear muscle reflexes. Special auditory tests, such as the auditory brainstem response test which measures the brain's responses to sound and x-rays, such as the MRI scan, can serve to confirm the diagnosis or atleast rule out some possible causes for the symptoms..*(cited in http://www.hearing center.com/info-files/sudden loss.html)*

Treatment of sudden hearing loss varies, depending on the suspected cause. The patient may be given medicine to increase circulation of the inner ear and to reduce excessive fluid build up in the middle or inner ear. Carbogen therapy, which increases the amount of oxygen to the inner ear may be started if there is a suspected circulation problem of the inner ear. A specific regimen is indicated if there is fluid build up in the middle or inner ear and surgery may be required for an inner ear tumor or a leak of inner ear fluid. The patient may be advised to stop the use of birth control pills, smoking, drugs, alcohol, caffeine, or to restrict the amount of strenuous activity, as well as watch the intake of fats and cholesterol.
.(cited in http://www.hearing center.com/info-files/sudden loss.html)

In summary, any one who notices any sudden diminution in hearing, regardless if there are any accompanying symptoms (fullness in the ear ringing or dizziness) should be seen by their ear, nose and thorat doctor within 24-28 hours.
.(cited in http://www.hearing center.com/info-files/sudden loss.html)

Recovery of the hearing loss depends not only upon the early diagnosis of the cause of the hearing loss, but also as the early initiation of appropriate treatment.

It is important for us to have an update of the review of literature available on "sudden deafness" for facilitating better understanding of this topic. As the literature reviewed with regard to this topic has been done till 1988 but not later, the main aim of this project work is to update the literature available in this area for ten years i.e. from 1988-1998 from various resources available. It is hoped that this project work will serve the purpose for which it is initiated. A glossary is presented at the end of the project to facilitate better understanding of key words used in this review.

INCIDENCE OF SUDDEN DEAFNESS

The incidence of Idiopathic sudden sensorineural hearing loss is estimated to be approximately 8 to 20% per 10,000 persons per year, although incidence may be even higher because some patients who recover spontaneously from sudden sensorineural hearing loss may not seek medical attention, (cited in http://www/utmb.edu/oto/Grnds.dir/ sudden deaf.htm).

Sudden deafness occurs in about one in every 5000 people every year. (cited in http://www.weizmann.ac.il/deaf-info/ suddendeafness.html).

The frequency of sudden deafness appears to increase with advancing age ranging from 4.7 per 10,000 persons 20-30 years of age to 15.8 per 100,000 persons 50-60 years of age. The mean overall age for sudden sensorineural hearing loss ranges from 46-49 years of age. The incidence of bilateral involvement is quite variable (1 -80%). There is no predilection to sex, and sudden sensorineural hearing loss does not occur in a seasonal pattern (cited in http://www/utmb.edu/oto/ Grnds.dir/sudden deaf.htm).

Non Otological Surgery Associated Hearing Loss (NOSAHL) was first reported by Jaffe (1967). Since then there have been sporadic reports, the majority of which (about 70%) have followed cardiopulmonary bypass surgery (Journeux and Greenhalgh, 1990). According to Erikson et al. (1992) sensorineural loss of sudden onset may be the presenting symptom in upto 14% of patients with acoustic neuroma.

Yanagihara and Asai (1993) state that in their series of 111 patients operated for acoustic neuroma from 1972 to 1990, 21 (i.e. 18.9%) had sudden hearing loss.

Moffat et al. (1994) in their study of 284 patients with vestibular schwannoma stated that 12% presented with suddensensorineural hearing loss and that 29.5% of those presenting with sudden deafness had total hearing loss.

Luxford and Saunders (1996) in their review of 823 sudden hearing loss patients from 1989 to 1993 state that only 1.7% had sudden bilateral hearing loss.

Hughes et al. (1996) in their review of literature state that approximately 4000 new cases of sudden hearing loss occur annually in the United States and 15,000 annually worldwide accounting for approximately 1 % of all cases of sensorineural hearing loss. In then-review of literature, they state that the incidence of sensorineural hearing loss increases with age, but there is no consistent sexual predominance or geographic clustering and that the disorder may be cyclic or seasonal.

Sudden hearing loss in patients with large vestibular aqueduct (VA) has been pointed out by several investigators since 1984. The incidence of sudden hearing loss in patients with large *VA* was relatively rare in many studies:l 9.1% (Emmett et al. 1988), 16.7% (Arcand et al. 1991), 1.8% (Jackler et al. 1989). However, a study conducted by Okumura and Takashaki (1998) observed as frequently as 60.9% of patients with large vestibular aqueduct presenting with sudden deafness.

CAUSES OF SUDDEN DEAFNESS

Since DeKleyn published the first group of patients with "idiopathic" sudden hearing loss in 1944, numerous clinical and laboratory investigations have attempted to identify the cause of this disorder, which in fact has numerous possible causes. Unfortunately, there is no better understanding of the etiology of sudden sensorineural hearing loss than there was one or two decades ago. (cited in http:// www.ummah.net/software/softbase/sudden.txt)

(Three hypothetical etiologies had been put-forth to explain the causes of sudden deafness. They are (i) viral infections (ii) vascular causes and (iii) labyrinthine membrane rupture Immune mediated inner ear diseases have been added to the triad of viral infection, vascular disorders and membrane rupture, (cited in http://www.ummah.net/ software/softbase/sudden.txt).

(Although viral infection of the inner ear is an attractive theoretical explanation for sudden hearing loss, the supporting evidence is not conclusive and consists of four types :

i) Coincident symptoms suggesting coincident viral infection .
ii) Serological data showing coincident viral infection.
iii) Histopathological studies showing similarities between known viral infections and cases of sudden hearing loss.
iv) Extrapolations from the degeneration of the inner ear seen in viral disease such as the prenatal rubella syndrome. (cited in http ://www. ummah.net/software/softbase/sudden.txt).

Uncontrolled studies from Van Dischoeck, Jaffe and Schuknecht in 1950s and 1960s reported "flu-like" symptoms in 20-60% of patients suffering idiopathic sudden sensorineural hearing loss. However a study by Rowson and Hinchcliffe in 1975 found the same symptoms of upper respiratory tract infection to be present in approximately 40% of the general population. Veltri et al. looked at 77 unmatched patients with sudden sensorineural hearing loss and found a conversion rate of 65%. Multiple viruses were implicated including influenza A and B, rubela, rubella, mumps, herpes simplex and cytomegalovirus. Wilson and coworkers studied 122 patients, 63% of whom seroconverted compared with 40% of controls. Other studies from Morrison and Booth as well as Bobison and Hinchcliffe failed to demonstrate a viral titre rise in sudden hearing loss patients. (The most consistent evidence supporting viral etiology is the histopathologic findings of varying degrees of atrophy of the organ of corti", stria vascularis and tectorial membrane with variable loss of the neural population as shown by Schuknecht and others. Perhaps the strongest evidence for the involvement of viruses in hearing loss comes from the use of immnunoflourescent antigen studies such as those performed by Davis and Johnson, demonstrating the ability of rubeola and mumps to infect the inner ears of animal models?) (cited in http://www. ummah.net/software/softbase/sudden.txt).

A vascular cause for sudden hearing loss is attractive because it is logically consistent with immediate onset of symptoms and because there are established models of acute hearing loss secondary to vascular occlusion in hypercoagulable states such as leukemia and sickle cell disease. However, most of the experimental and clinical evidence casts serious doubts at vascular etiology. (cited in http://www.urnmah.net/ software/softbase/sudden.txt).

One would expect that older patients with known peripheral vascular disease would be the ones most often afflicted, but infact, most patients are younger and have no stigmata of the systemic vascular disease. Perlman and other investigators have demonstrated permanent loss of the cochlear microphonic and action potential occurs after only 30 minutes of ischemia. The histopathology of ears affected by sudden hearing loss also differs significantly from temporal bones examined after experimental vascular occlusion. Permanent obstruction leads to marked degeneration of the neurons followed by fibrous and osseous proliferation within the inner ear, changes that are not seen typically in sudden sensorineural hearing loss. (cited in http://www.ummah.net/ software/softbase/sudden.txt)

Intracochlear membrane breaks were proposed as a cause of sudden hearing loss by Simmons in 1968, but the evidence is only coincidental.

Schuknecht and Denovan studied 12 temporal bones of patients afflicted with sudden sensorineural hearing loss and found no evidence of Reissner's or basilar membrane rupture, (cited in http:// www. ummah. net/software/softbase/sudden. txt)

There are several autoimmune diseases that are epidemiologically associated with sudden sensorineural hearing loss, including Cogan's Syndrome, systemic lupus erythematosus, and temporal arteritis. Some of the most convincing evidence has come out of the University of Tennesse by Yoo and coworkers who showed that monoclonal antibodies specific for type II collagen can activate an immunologic response in the inner ear of the rate leading to sensorineural hearing loss as

documented by ABR testing. They were able to demonstrate perivascular inflammation and fibrosis and degeneration of the spiral ganglion on histopathology and using immunofluorescent techniques they saw immune complexes in the otic capsule, (cited in http://www.ummah.net/software/softbase/sudden.txt)

Apart from the four main theoretical hypothesis put-forth to explain the aetiology of sudden deafness, there have been various etologies reported for sudden deafness.

Franklin et al. (1989) report of two cases of multiple sclerosis where sudden sensorineural hearing loss was the initial presenting manifestation in one patient and an exacerbation of the longstanding disease in another. Multiple sclerosis is known to affect the myelin of the auditory pathway resulting in acute hearing loss. While the identification of central auditory pathway abnormalities in multiple sclerosis is a frequent finding, sudden sensorineural hearing loss *is* an uncommon presenting symptom in this disease.) Auditory system involvement is usually in conjunction with associated disturbances in the brainstem, thereby making the diagnosis of this demyelinating process easier. Isolated sensorineural hearing loss can be a rare initial manifestation of multiple sclerosis, but the cause may remain obscure until either additional neurologic deficits develop or clinically silent plaques within the central white matter are detected on roentgenographic imaging.

Drulovic, et al. (1994) also report of two patients who presented with sudden deafness and tinnitus as the initial symptoms for multiple

sclerosis. Two more cases of sudden deafness due to multiple sclerosis have been reported by Marangos in 1996.

The relationship between sudden sensorineural hearing loss and trauma has traditionally thought to be due to the presence of a perilymphatic fistula in which an abnormal communication exists between the middle and inner ears. Goodhill (1971) introduced the concept of labyrinthine membrane rupture and proposed two possible mechanisms of injury in a perilymphatic fistula : the implosive and explosive results. The implosive route results from transmission of increased middle ear pressures due to valsalva maneuvers to the perilymphatic space through the oval and round windows. The explosive route on the other hand, results from transmission of increased intracranial cerebrospinal fluid pressure to the perilymph via either the cochlear acqueduct or the internal auditory canal.

Shelton and Lusted in 1989 proposed an animal model to explain the role of perilymphatic fistula in sudden hearing loss. The perilymphatic fistulas are usually either tears in the ligamentous attachments of the stapedial footplate to the bone of the oval window or in the round window membrane. The causes of a peripymphatic fistula are multiple including stapedectomy, head or ear trauma, diving, flying, surgery for chronic otitis media, valsalva like exertion, general anaesthesia, acoustic trauma and congenital perilymphatic fistula.

Jackler (1989) summarised four possible etiologic mechanisms for sudden hearing loss in the enlarged vestibular acqueduct syndrome : three of these attempt to explain how minor head trauma may precipitate

sudden hearing loss and the last theory attempts to explain the progressive hearing loss seen.

The "Reflux theory" of Levenson (1989) hypothesises that mild head trauma may compress the dural envelope of the endolymphatic sac, allowing the back wash of high protein hyperosmolar fluid into the endolymphatic sac through the abnormally patient vestibular acqueduct into the cochlea.

The "Intracochlear Membrane Rupture" theory of Jackler suggests that congenital weaknesses may exist between the membraneous cochlear partition. Thus, traumatic intracochlear membrane ruptures of either the basilar membrane or Reissner's membrane may result in the mixing of the perilymph and endolymph producing stepwise sudden loss of hearing.

The final theory that relates trauma to sudden SHL in the enlarged vestibular acqueduct syndrome is the "perilymphatic fistula" theory. Belenky (1953) was the first to describe a post traumatic round window fistula in association with this syndrome.

The fourth theory proposed by Jackler is the "Endolymphatic Hydrops" theory in which the endolymphatic sac is thought to be dysfunctional, resulting in the accumulation of excess volumes of endolymph and producing progressive SHL.

Kou and McDonald (1998) reported that a perilymphatic fistula is not the only mechanism for traumatic SHL, and the 'Reflux' theory

of Levenson as wellas the 'Intracochlear Membrane Rupture' theory of Jackler may be just as important in the pathogenesis of the hearing loss.

Timon et al. (1989) report of two cases of HIV positive patients with sudden sensorineural hearing loss as the presentation of the HIV infection.

O'Keeffe and Maw (1991) report of the first case in literature with sudden onset bilateral hearing loss due to sickle cell disease which is a haemolytic anaemia and is most prevalent intropical and subtropical regions. The disorder is inherited along mendelian lines and may be associated with other haemoglobinopathies, most commonly with thalassaemia trait. Symptoms of anaemia are usually mild because haemoglobins (HbS) readily releases oxygen to the tissues but the disease is punctuated by painful cries induced by episodes of infection, dehydration or deoxygenation. At such times, sickle cells obstruct small vessels by sludging and thus cause ischaemia and tissue necrosis.

Katholm et al. (1991) report of an 18 year old woman, suffering from acute acquired toxoplasmosis, who had experienced sudden deafness and a total loss of vestibular function first in right ear and three months later also in the left. Toxoplasmosis is an infectious disease caused by a protozoan, Toxoplasma gondii. The cat is the principal host and may excrete infectious oocysts. Infection in man occurs after ingestion of contaminated undercoaked meat or by contact with infectious animals (Krick and Remington, 1978). The ingested parasites invade the epithelium of the intestine, multiply in the mesentric lymph nodes.

Hall et al. (1991) reported that five out of twelve patients who reported of sudden sensorineural deafness had red blood cell deformability, thus suggesting that real red blood cell deformability may be another etiology for sudden deafness.

Yanagihara and Asai (1993) reported that 21 (18.9%) of 111 patients operated for acoustic neuroma from 1972 to 1990 had sudden hearing loss. They reported that even a small tumor has the potential to produce sudden hearing loss and the recognition of sudden hearing loss as an initial symptom of acoustic tumor is essential to detect small acoustic neuroma.

In about 10% of patients with an acoustic neuroma, the hearing-impairment is reported to have come on suddenly. This does not mean that the patient first happened to notice the loss on a specific occasion. Because of the relatively rarity of acoustic neuroma it would be incorrect to imply that an individual with a sudden hearing loss is likely to have one.

Pringle et al. (1993) report of a case of sudden sensorineural hearing loss due to metastatic prostratic carcinoma in the temporal bone. Despite the fact that carcinoma of the prostrate is a very common neoplasm and is well known for metastasizing for to the bone, this case presented is the first case history in English literature of a prostatic metastasis to the temporal bone causing sudden sensorineural hearing loss.

Alles and Pye (1993) exposed the cochlea of pigmented guinea pigs to an 8 KHz puretone at 116 dB SPL for 1 hour and or 50 mg. Kg day of gentamicin for 10 consecutive days and repeated after an interval of 3 weeks. Hair cell loss was found to have occurred in the contralateral cochlea following the sound exposure alone.

The following is a list of common drugs that can be cochleo toxic :

Class	Drug	Proprietary names
Non-steroidal anti-inflammatory	Salicylates	Alkaseltzer, Antoin, Benoral, Claradin, Codis, Hypon, Labofrin Levius, Unadox, Paynocil, Sofapryn, Veganin.
Beta Blockers	Propanolol	Angilol, Apsolol, Bekolol, Inderal
	Acebulotol	Sectral
	Atenolol	Tenormin
	Labitolothydro-chloride .	Trandate
	Metoprololtartrate	Betaloc, Loprisor
	Nadaloll	Corgard
	Oxprendol - hydrochloride	Apsolox, Trasicor
	Pendolol	Visken
	Sotalol-hydrochloride	Beta cardone, sotacor
	Timolol maleate	Betin, Blocadren.
Antibiotics	Gentamicin	Cedomycin, Garamycin, Genticin.
	Amikacin	Amikin
	Framycetin-Sulphate	Soframycin
	Kanamycin	Kannasyn, Kantren
	Neomycin	Mycifradin, Nivemycin
	Netilmicin	Netillin
	Trobamycin	Nebcin

Class	Drug	Proprietary Names
Loop diuretics	Frusemide	Dryptal, Frusetic Frusid, Lasix
	Bumetanide	Burinex
	Eltracrynic acid	Edecrin
Metals	Cisplatinum	Neoplatin

Hakan Einer and Lilian Tengborn in 1994 studied tie role of pathologic homeostatic mechanisms as a possible cause of sudden sensorineural hearing loss. They studied 32 consecutive patients with sudden hearing loss and 28 healthy individuals(Control groups). Results indicated that25 of the patients had some kind of aberration of specific homeostasis parameters; seven had an increase in the activity of plasmogen activator inhibitor 1 (a glycoprotein associated with diminished fibrinolysis) compared with the control group. Increased plasminogen activator inhibitor levels were most frequently observed among the patients who were overweight. Seven of the oldest patients had an increase in D-dimers which is a dehydration product of fibrin and most of their patients had a history of cardiovascular disease. Although isolated aberrations in the homeostatic pathway were observed, they concluded that pathologic homeostasis does not have decisive importance for the pathogenesis of sudden deafness.

Moffat et al. (1994) reported that 12% ofa series of 284patients with vestibular schwannoma presented with sudden deafness. Although the number of patients with sudden deafness in this series were too small to reach significance, on the basis of the clinical correlation of vestibular schwannoma morphology, it is possible to postulate that

compression of the vasculature within the bony internal auditory canal by a laterally arising tumor may be the aetiological factor and may be more likely to occur than in more medially arising tumors.

Galvez et al. (1994) described the case of a female patient who presented with sudden deafness as the first symptom of a cerebellar tumour which was not localised strictly in the CP angle and did not show direct compression on the extrabulbar portion of the VIII cranial nerve. The clinical picture contained a number of signs and symptoms typical of cerebellar involvement, surgical intervention restored the hearing and caused the symptoms to disappear.

Ohinata et al. (1994) suggest that blood/plasma viscosity may be involved in the etiology and prognosis of sudden deafness and hence point to the importance of measuring blood and plasma viscosity in patients with sudden deafness. They measured blood viscosity and plasma viscosity in 51 patients with sudden deafness and 70 controls with normal hearing. Their results suggested that many patients with sudden deafness have increased blood viscosity and plasma viscosity and that this increase may play a significant role in the etiology of sudden deafness.

Okumura et al. (1995) examined 181 patients (327 ears) with sensorineural hearing loss of unknown etiology and 25 people (50 ears) with normal hearing by high resolution computed tomography, the image of the large vestibular acqueduct (VA) was defined as being a visible large aperture (>4 mm) and small distance between vestibule and traceable part of the vestibular acqueduct nearest to the vestibule (>1

mm). The large vestibular acqueduct was found in 13 patients (23 ears; 7.0%); it was relatively frequent following hypoplastic cochlea(33 ears, 10.1%) in all the inner ear anomalies. In patients with large vestibular acqueduct, high frequency hearing was affected more than low frequency and history of sudden hearing loss was observed frequently. 61% of ears with large VA was found to be triggered by characteristic episodes such as minor head trauma etc.

Ravi and Henderson (1996) report of a diabetic presenting to an otolaryngologist with sudden deafness of six days duration as the only symptom. This is the first reported case in literature of any patient presenting with sudden deafness as the only symptom of diabetes mellitus.

Hughes et al. (1996) give a partial list of causes of sudden sensorineural hearing loss. They are as follows :

1. Infectious causes : These include Meningococcalmeningitis, herpes virus (simplex, zoster and varicella), mumps, AIDS, mononucleosis, lassa fever, mycoplasma, cryptococcal meningitis, toxoplasmosis, syphilis, cytomegalo virus, rubeola, rubella and human spumareto virus.

2. Traumatic causes : These include - Perilymph fistula, inner ear decompression sickness, temporal bone fracture, inner ear consussion, otologic surgery, surgical complications of non-otologic surgery.

3. Neoplastic causes : These include acoustic neuroma, leukemia, myeloma, metastasis to internal auditory canal, meningeal carcinomatosis, contralateral deafness after acoustic neuroma surgery.

4. Immunologic causes : including primary immune inner ear disease, temporal arteritis, Wegener's granulomatosis, cogan's syndrome, polyarteritis nodosa, delayed contralateral endolymphatic hydrops.

5. **Toxic causes** : include snake bite, ototoxicity etc.

6. **Circulatory causes** : including vascular disease/alteration of microcirculation, vascular disease associated with mitochondriopathy, vertebrobasilar insufficiency, redblood cell deformability, sickle cell disease, anamolous carotid artery and cardiopulmonary bypass.

7. **Neurologic causes** : include multiple sclerosis and focal pontine ischaemia.

8. **Metabolic causes** : include thyrotoxic hypokalemia, disturbances of iron metabolism, diabetes mellitus and renal failure/dialysis.

9. **Other causes:** These include meniere's disease, pseudohypoacusis, neurosarciodosis, cyclosporin treated renal transplantation, dental surgery, hyperostosis cranialis interna, genetic predisposition and stress.

Cox and Sargent (1997) report three cases of sudden sensorineural hearing loss following nonotologic surgery in which cardiopulmonary bypass (CPB) surgery was not involved.

Cruz and Bance (1998) described a case of permanent bilateral severe hearing loss following metatarsal pinning in a patient with pre-existing non-operated otosclerosis (nonotologic surgery).

Brewis et al. (1997) give the first case report in literature presenting with sudden sensorineural hearing loss in association with Chlamydia psittaci infection. Chlamydiaceae are a group of agents with mixed alligiance; they are obligate intracellular pathogens like viruses but contain both DNA and RNA like bacteria. The genus Chlamydia contains three species : C.psittaci; C.pneumoniae, C.aschomatis, all of which can cause disease in human. C.psittaci has an avian reservoir and cases of penumonia following exposure to infected birds are well recognised. C.psittaci infection has been reported in association with a case of Cogan's syndrome, a syndrome characterised by fluctuating sensorineural hearing loss, vertigo, uveites and keratitis (Darougar et al. 1978).

Garcia et al. (1997) reported significant abnormalities in the subpopulations of lymphocytes in patients with sudden hearing loss suggesting the existence of immune mediated responses in the inner ear as possible etiopathogenic factors in this entity.

Suzuki et al. (1997) reported four patients with bilateral sudden hearing loss related to gastric adenocarcinoma.

Kou and MacDonald (1998) studied cases at the Hospital for Sick Children, Toronto between the years 1980 and 1995. High resolution CT scans of the temporal bones with bone algorithms and coronal/axial view were performed on all children presenting with sudden hearing loss after 1988. Of the 12 children studied, 9 had high resolution CT scan, revealing a 33% incidence each of inner ear malformation, temporal bone fractures and other miscellaneous CT findings.

Takasaki et al. (1998) in their study of serum antibodies to human herpes virus 7 (HHV-7), human herpes virus 6 (HHV-6) and cytomegalovirus (CMV) in patients with idiopathic facial nerve palsy or sudden deafness found that the antibody titres to CMV, HHV-6 and HMV-7 did not increase in majority of the patients with Bell's palsy and sudden deafness and concluded that CMV, HHV-6 and HHV 7 are not the direct cause of Bell's palsy and sudden deafness in most patients.

Sudden hearing loss may either be unilateral or bilateral. Some of the recognised causes for sudden **unilateral** hearing loss include :

* Herpes zoster.
* Typical viruses such as influenza
* Mumps, syphilis (congenitalor acquired).
* Membrane rupture/fistulas in the inner ear .
* Vascular disorders.
* Acoustic Neuromas
* Head/acoustic trauma
* Meniere's disease.
* Unknown causes. (cited in inhttp://www.aos_jax.com/sudd.hl.htm)

Recognized causes for sudden **bilateral** hearing loss :

* Systemic infections
* Meningitis
* Scarlet fever
* Typhoid fever
* Measles.
* Tuberculosis
* Syphilis .
* Autoimmune disorders
* Ototoxic medications (generally used in life threatening situations)
* Multiple sclerosis .
* Unknown causes (cited in inhttp://www.aos_jax.com/sudd.hl.htm) .

Other causes of Sudden hearing loss include(Browning, G. G.(1986)).

1. Barotrauma : Barotrauma, with rupture of the round window membrane, can occur after an aeroplane flight or after underwater swimming or diving. The mechanism whereby this can occur is worth explaining.

The external auditory canal, middle ear and inner ear can be considered to be three individual compartments, separated from each other by the tympanic membrane and the round and oval window membranes. The pressure in the external auditory canal is that of the environment. The pressure in the middle ear is that of the environment, unless the eustachian tube is malfunctioning. The pressure in the inner ear is essentially that of the environment except that the perilymph pressure can be transiently increased by an action which raises the intracranial pressure such as coughing or sneezing. This is then transmitted from the CSF to the inner ear via the cochlear acqueduct. A pressure difference between any two of the three compartments can cause the separating membranes to tear.

During aeroplane ascent, the atmopheric pressure and that of the external auditory canal and inner ear can fall by as much as 1 atmosphere (-760 mm Hg). Even if the eustachian tube is not functioning, the middle ear air can escape down the eustachian tube so the pressure differential usually readily equates. During an aeroplane ascent it is thus uncommon to have otalgia due to differential pressure across the tympanic membrane.

The main problem arises during descent when the pressure in the external auditory canal and inner ear rises back to atmospheric pressure. The middle ear is then at a relative, negative pressure of-760 mm Hg to the external auditory canal and the inner ear. It can be difficult even for a normally functioning eustachian tube to open up against this pressure differential and it is obviously more so if the tube is oedematous because of an upper respiratory tract infection. So otalgia is common during an aeroplane descent and on occasions the tympanic membrane can rupture. Alternatively, the round window membrane may rupture and perilymph leak into the middle ear and this is considered more likely to occur if there is an additional increase in inner ear pressure occasioned by a cough or a sneeze, but this only increases the pressure by 1.5 mmHg.

Similar pressure differentials occur during diving. Thirty feet (10 metres) underwater is equivalent to an additional one atmosphere (760 mm) of pressure. Against this is the fact that divers are taught how to perform a valsalva manoeuvre and most do this regularly and effectively. If this is ineffective there is, in theory, the possibility that will make matters worse by increasing intracranial pressure. The pressure that this achieves has not been recorded but is unlikely to make a material difference.

Deep sea divers are an interesting group to study because there are several reasons why they might have hearing problems after a dive. Serous otitis media can occur, due to rebound vasodilatation of the middle ear and eustachian tube mucosas, but is not too difficult to diagnose. Rupture of the tympanic membrane should also present little diagnostic

difficulties. What can sometimes be difficult to differentiate is between decompression sickness and barotrauma causing a round window rupture. Decompression sickness is often associated with disequilibrium but there should be no nystagmus. The vast majority of divers with presumed round window ruptures have vertigo and nystagmus at the time of the incident which would make one wary of making such a diagnosis in the absence of vertigo. Oval window rupture has rarely been reported in divers which must throw some doubt on this as a frequent entity except where there has been previous oval window surgery.

In individuals with a sudden sensorineural hearing loss recent aeroplane flights and diving should be inquired about. If the round window has ruptured, vertigo and nystagmus will almost invariably be present and if this is not the case then the diagnosis must be seriously doubted. Reports of round window ruptures following coughing, sneezing or straining must be treated sceptically considering that the round window membrane is a closely knit fibrous structure (Normura et al. 1983) and coughing, sneezing or straining only raises the intracranial pressure by 1.5 mm Hg (150 mm H_2O).

2. Ear Surgery : Though it might not be admitted, ear surgery is probably the commonest cause of a sudden hearing loss after wax and secretory otitis media. Indeed a total hearing impairment can follow any otological operation, the degree of risk depending on the surgeon's expertise, which operation is being performed and whether a middle ear infection coexists or subsequently occurs.

The risk is highest at the time of surgery, especially if the oval window has been opened or if a semicircular canal fistula was present or created, but the loss can occur at any time.

In the immediate postoperative period, inner ear damage should be suspected if there is vertigo and spontaneous nystagmus, but it is difficult to clinically assess the hearing because of ear dressings. Should inner ear damage be suspected, a decision has to be made as to whether the ear should be reexplored. In general, most otologists would continue to observe the patient for several days but others might reexplore in the hope of sealing a perilymph leak, especially following a stapedectomy, but the risk of causing yet further inner ear damage is considerable.

3. **Head Injury** : It is estimated that in the UK there are nearly, million attendances at hospital or general practice with a head injury per year and in a proportion, the auditory system will be affected. The external auditory canal may be filled with blood as might the middle ear, the tympanic membrane might be ruptured, the ossicular chain may be disrupted by the blow, the temporal bone may be fractured, the auditory nerve may be stretched in its canal and the brainstem and cerebrum may be damaged directly. The incidence of each has never been fully ascertained but permanent sensorineural impairments caused by temporal bone fractures are perhaps commoner than realised. This is because radiology is not particularly successful in detecting basal skull fractures and correspondingly an otoscopic examination should be performed in all individuals with a head injury to ensure that temporal bone fractures are not missed. They are most usually evident by blood or cerebrospinal fluid (CSF) being present in the external auditory canal.

Five percent of all minor head injuries with a short period of post-traumatic amnesia (Browning and Swan, 1982) and 25 percent of all severe head injuries (Browning, unpublished observation) will have otoscopic evidence of a temporal bone fracture and the majority of these will not have been detected on straight or tomographic radiology of the skull.

Individuals who do not have a temporal bone fracture are unlikely to have damaged the peripheral auditory system, but a considerable proportion will have central auditory problems and disequilibrium due to whiplash injuries to the brainstem.

A rarer variant of a head injury is a blast injury to the ear, most common these days as a result of explosions in civil disorders. This usually results in a rupture of the tympanic membrane, but inner ear damage can also result.

4. Noise : The exposition to acute acoustic trauma (gunfire injuries and explosion) for instances can cause severe and sudden hearing loss. The cause is partly a direct and mechanical one, due to bleeding and partly an indirect metabolic effect on the microcirculation causing partially reversible damage to the sensory cells of the organ of corti.

5. Psychogenic : Sudden hearing loses with a psychogenic basis are usually bilateral and therefore easily distinguished from the losses which are almost invariably unilateral.

6. Secretory otitis media : Secretory otitis media following an upper respiratory tract infection is common and usually cause sudden hearing loss.

7. **Syphilis** : Very rarely, early acquired syphilis is complicated by meningitis and this usually presents with various neurological symptoms and signs. The auditory nerve may be involved and there are reports of deafness being the sole presenting symptom. Because it is so important to treat syphilitic meningitis there is an argument for routinely performing syphilis serology in individuals with a sudden sensorineural hearing-impairment.

8. Wax : The impaction of wax against the tympanic membrane with a cotton bud or the swelling of it with water to occlude the canal are common causes of a sudden hearing loss which are easy to diagnose and remedy.

SYMPTOMS

The most outstanding feature of sudden deafness is its sudden onset of symptoms. The symptoms can be classified into three main categories (i) Deafness (ii) Tinnitus (iii) Vertigo.[Snow, J.B(1973)]

(i) **Deafness**

A great deal of variation occurs in the evolution of symptoms of sudden deafiiess. The deafiiess may begin instantaneously, and when the onset is so sudden, it may be accompanied by the sensation of a loud sound in the affected ear. More often the hearing loss develops over the course of an hour, a day or several days. Frequently the hearing loss is first noticed by the patient on awaking in the morning. Some patients are awakened from sleep by the associated tinnitus. If the loss is bilateral, it will be noted promptly once interpersonal communication is hindered. On the other hand, unilateral losses may escape the patient's detection until some specific test of the affected ear, such as use of the telephone, occurs. Dating of the onset is often difficult in children and in adults, as well. Usually the hearing loss draws the patient's attention promptly regardless of the presence/absence of tinnitus. The patient is often aware of the profound loss of discrimination that is initially present. Unusual sensitivity to intense sound and diplacusis are rare. As a rule, difficulty in localizing sound is experienced.

(ii) **Tinnitus**

Roughly 70% of patients with sudden deafiiess experience tinnitus of varying degrees sometime during their illness. The tinnitus

may precede the hearing loss by several hours; it usually subsides with in one month but it may persist and even outlast the deafness. It usually has a roaring quality.

(iii) Vertigo

There is considerable variation in the incidence of vertigo from one series of patients to another. In general, some 40% of the patients with sudden deafness have mild/transient vertigo which lasts for four to seven days. Lesser degree of vertigo may then persist for up to six weeks. Nausea and vomiting are usually associated with severe vertigo.

A sensation of pressure in the affected ear is experienced *by* many of these patients.

Headache is occasionally encountered, and symptoms of viral upper respiratory tract infections occur as frequently as in 25% of the patients in some series. Fever, usually of mild degree, may be present. Generally, however, the patient feels perfectly well except for the loss of hearing and tinnitus.

Usually the otoscopic examination is normal, but serous otitis media is occasionally observed and may add a conductive component to the loss of hearing.

Franklin et al. (1989) report of two cases of multiple sclerosis with sudden sensorineural hearing loss. Low pitched tinnitus and decreased auditory acuity was noticed in one patient on awakening in

the morning while the other had no associated vertigo or tinnitus associated with reduced auditory acuity which occurred suddenly.

Nakashima et al. (1992) in their report of measurement of cochlear blood flow during tympanotomy in four patients With sudden deafness report that out of four patients, three reported of tinnitus. Vertigo was present in all four patients. Flat perceptive loss was noticed in the fourth patient. The audiogram showed slight hearing loss with levels within 25 dB with an exception of a 35 dB reading at 8 KHz.

Nakashima and Yanagita (1993) in their review of 1313 patients who presented within two weeks after the onset of unilateral sudden deafness between 1972 to 1990 report that 30% of the patients had accompanying vertigo and this occurred frequently in patients with severe hearing loss in high tone frequencies.

Okumura et al. (1993) in their study of 98 patients with idiopathic sudden deafness report that 79% of the patients complained of tinnitus, 23% of vertigo or some form of unsteadiness.

Shairaishi et al. (1993) reported in their study of 98 patients with 'idiopathic' sudden deafness that 79% of the patients complained of tinnitus and 23% of vertigo or some form of unsteadiness.

Drulovic et al. (1994) report of two patients with "Multiple sclerosis" with sudden deafness and tinnitus as the initial symptoms of multiple sclerosis. No vomiting or vertigo was reported in both the patients.

Kanda et al. (1995) in their report of sudden hearing loss associated with "Interferon" reported that auditory disability (tinnitus/ hearing loss) occurred in 32 patients (43.8%) during Interferon therapy, among whom audiometry documented sensori-neural hearing loss in 27 cases (i.e. 36.9%). 17(46.8%) of the 35 patients receiving Interferon-beta (IFN - β) had auditory disability including hearing loss in 13 cases (37.1%) and 15 (39.5%) of 38 patients receiving Interferon - alpha (IFN - α suffered from auditory disability. Not much influences on symptoms on the influences of IFN-α and β was seen. According to the authors, auditory disability frequently developed in the later stages of treatment

Shitara et al. (1996) in their report of a series of 127 patients with idiopathic bilateral sensorineural hearing loss report that rapid progression of idiopathic bilateral sensorineural hearing loss was accompanied by dizziness in two patients. The frequency of appearance of dizziness associated with rapid progression of idiopathic bilateral sensorineural hearing loss was 25%, being lower than in sudden deafness.

Suzuki et al. (1997) in their report of four patients with gastric adenocarcinoma presenting with bilateral sudden hearing loss report that tinnitus was noticed in two patients, vertigo in three patients, nystagmus in two patients, otalgia in one patient and facial nerve paralysis was noticed in one patient.

Cox and Sargent (1997) in their report of three patients of sudden sensorineural hearing loss following non-otologic surgery in which cardiopulmonary bypass surgery was not involved,report that all the

three cases had a complaint of aural fullness and hearing loss and light headedness was reported by one of the three patients.

Stokroos et al. (1998) in their study of 43 Dutch and Flemish Idiopathic sudden sensorineural hearing loss patients between 1994 and 1996 report that -

a) 36/43 patients (84%) experienced tinnitus, either preceeding, coinciding with or following hearing loss. Tinnitus persisted for six months in 24/43 (56% of patients).

b) Twelve months after hearing loss, 17/43 (40% of patients still experienced tinnitus. Severity of tinnitus did not differ significantly across categories (mild, moderate and severe).

c) A pressure sensation on the affected ear was present in 21/43 patients (i.e. 49%). In six cases, this sensation preceeded the hearing loss, in seven cases, it accompanied the hearing loss and in eight cases, it was noticed following the hearing loss.

In six cases, pressure was judged to be mild, twelve cases it was moderate and in three it was severe.

d) Hearing loss was accompanied by a sense of disequilibrium or vertigo in 20/43 cases (i.e. 47%). In 11 cases, this sensation became apparent after the hearing loss occurred, in four it accompanied hearing loss and in two it preceeded hearing loss.

HISTOLOGICAL STUDIES

There have been very few histological studies conducted on patients with sudden deafness.

Early studies by Schuknecht in 1962 and Beal et al. in 1967 represent the final stages of the processes in patients in whom spontaneous recovery does not occur. Their findings can be considered as the end result of the most virulent infections and do not represent the outcome in all cases. They report that the organ of corti is often missing in the basal turns, and the individual hair cells tend to be missing at higher turns. Ganglion cell populations are decreased at the basal turn but are more normal towards the apex. The stria vascularis tends to be atrophic. The tectorial membrane is often atrophic, rolled up and ensheathed in a syncitium of cells on the limbus. Reissner's membrane may be collapsed and adherent to the basilar membrane. The saccule is often involved, but the utricle and semicircular canals usually escape severe damage.

Alford et al. (1965) and Suga et al. (1970) have studied experimental microembolization in animals and as in other forms of vascular occlusion, degeneration of the spiral ganglion cells to the apex of the cochlea and fibroblastic invasion and new bone formation in the cochlea were prominent.

Khetarpal (1989) investigated the temporal bone histopathology of sudden deafness patients with and without vertigo. He hypothesized that vertigo in sudden deafness was caused by

transmission of a change in inner ear fluid from the cochlea to the vestibular apparatus because he observed no direct relationship between the presence of vertigo and the damage to the vestibular apparatus.

Khetarpal et al. (1990) studied 22 temporal bone specimens from 18 patients who during life suffered a sudden partial/complete sensory-neural hearing loss. The cases were assigned to one of the following three diagnostic categories:

Group-1 : Idiopathic sudden sensorineural hearing loss with no history of an upper respiratory tract infection.

Group-2 : Idiopathic sudden sensorineural hearing loss with a history of upper respiratory tract infection preceeding or occurring, concurrently with the hearing loss.

Group-3 : Postnatal presumptive viral labyrinthitis, consisting of cases of profound hearing loss in one/both ears following an attack of measles, mumps or herpes zoster oticus.

The authors present a single case study under each group. This is as follows :

Case 1 (Group 1):

A 65 year old man who experienced sudden profound sensori-neural hearing loss in left ear that persisted until his death six years later. This was associated with nausea and vomiting that lasted for several hours.

Histological study of this patient revealed atrophic changes in the organ of corti with loss of pillar and hair cells. Total loss of organ of corti in the basal 15 mm was seen. There was moderately severe atrophy of the stria vascularis at various locations in the cochlea and partial loss of ganglion cells in the basal turn. The vestibular labyrinth was normal.

Case 2 (Group 2) :

A 39 year old woman who awakened with hearing loss and tinnitus in right ear. She had complained of a concurrent head cold and nasal stuffiness. No history of vertigo was reported. At the age of 59, the year of her death, audiometric tests showed a moderate sensory neural hearing loss on right and normal hearing on left. Tests of speech discrimination were not done.

Histological study, of this patient revealed a moderate loss of hair cells of cochlea with severe loss of hair cells in the saccule oft the right ear. The spiral ganglion cell population was normal. The organ of corti, cochlear neurons, vestibular sense organs and neurons of the left ear appeared normal.

Case 3 (Group 3) :

This patient was known to have been profoundly deaf following an illness diagnosed as measles at the age of four years. She died at 53 years of acute myocardial infarction.

Histological study showed severe loss of hair cells on the right and total loss on the left. There was a severe loss of stria vascularis bilaterally and a severe endolymph hydrops in right ear. A moderately severe loss of spiral ganglion cells was seen bilaterally that was maximally at the base. The saccular maculae showed patchy atrophy bilaterally, but it was more severe on the right. The utricular maculae was normal on the left and showed mild atrophy on the right.

Moore et al. (1997) studied the degenerative changes in the central auditory system in seven subjects with profound bilateral adult onset deafness. Each of the seven subjects became profoundly deaf bilaterally during adolescence or adulthood. Three of the subjects had inflammatory conditions (two cases of bacterial labyrinthitis secondary to meningitis one case of probable viral labyrinthitis). One subject had idiopathic progressive hearing loss, one lost hearing following removal of bilateral acoustic neuromas caused by "Neurofibromatosis 2" (NF 2) and the remaining two cases were diagnosed as adult onset "Scheibe degeneration" on the basis of postmortem temporal bone histopathology. The degree of transneuronal atrophy was determined by measuring cell size at three levels of the brainstem auditory pathway (Antroventral cochlear nucleus , medical superior olivary nucleus, and inferior colliculus). Within subjects, the relative degree of cell shrinkage was similar across all levels of the central pathway. Across subjects, the best neuronal preservation was seen in a case of viral labyrinthitis with one year of bilateral deafness and a near normal population of cochlear ganglion cells. Reduction in cell size was greatest in cases of bacterial

labyrinthitis or Scheibe degeneration with reduced populations of ganglion cells and longer periods of deafness. At the level of the cochlear nucleus, there was no consistent difference in cell size between the size stimulated by a functioning prosthetic device and the non-stimulated side.

Suzuki et al. (1997) studied six temporal bones (three patients) and a brain tissue sample (one patient) removed at a autopsy with bilateral sudden hearing loss related to Gastric adenocarcinomai According to the authors, the two mechanisms which might have caused bilateral sudden deafness in patients with adenocarcinoma is (i) metastasis to the internal auditory meatus damaging the corti's organ, or (2) inner ear haemorrhage damging cortit organ.

Case 1

A 54 year old man underwent a gastrectomy for treatment! of gastric adenocarcinoma Half a year later, he lost his hearing ability bilaterally three weeks before his death. He developed right facial nerve palsy two weeks before his death, and left facial nerve palsy one week before his death. He complained of headache and vertigo and horizontal nystagmus to the left was observed. He died of diffuse "Leptomeningeal carcinomatosis".

Temporal bone histopathological findings :

The most significant pathological finding was the massive infiltration of carcinomatous cells into the internal auditory meatus,

involving the VII and VIII cranial nerves. The auditory nerve was replaced by cancer cells. However, the organ of corti, saccule and utricle were well preserved. No marked histopathological differences were found between his right and left ears. There was a small amount of blood in the scala vestibuli.

Case 2

A 64 year old man underwent gastrectomy for treatment of Borrmann type II gastric-adenocarcinoma. Eight months later, he suddenly experienced tinnitus, otolgia, hearing loss and vertigo in his left ear. He lost his hearing ability progressively, resulting three weeks before his death, in profound bilateral hearing loss and canal paresis. Cerebrospinal fluid examination revealed dissemination of cancer cells in the subarachnoid space. Six weeks after his initial visit he died of "Leptomeningeal carcinomatosis".

Histopathological findings

Temporal bone dissection was not performed on his family's request. The brain was oedematous and histopathological examination revealed diffuse invasion of the arachnoid space by signet cells. Metastasis to the brain parenchyma was not observed.

Case 3

A 3 8 year old man without any past serious illness presented suddenly with nasal bleeding and pain in the hip joint. On admission,

RI scintigram revealed metastatic carcinoma of unknown origin in the left femur. His platelet count became extremely low (30,000) and he died of leukaemoid reaction three weeks later.

Thirteen days before his death, he suddenly developed severe hearing loss in the left ear, confirmed by puretone audiometry. The next day he became completely deaf in both ears and did not recover before his death. He complained of slight vertigo but nystagmus was not reported. No other symptoms such as facial nerve paralysis was observed.

Autopsy revealed the cause of the leukaemoid reaction to be bone metastasis of gastric carcinoma.

Temporal Bone Histopathological Findings

Right ear : Massive bleeding was observed both in the perilymphatic and endolymphatic spaces. Tectorial membranes were missing in most turns. The spiral ligament and modiolus were filled with blood. The vestibular organs were filled with blood.

Left ear **:** Blood had accumulated mainly in the perilymphatic space of the cochlea. The organ of corti, tectorial membrane and stria vascularis showed better preservation.

Case 4

A 46 year old man without any particular past medical history began experiencing back pain. Three weeks later, he developed right hearing loss without vertigo over a period of 12 hours. Audiometry revealed severe sensorineural right hearing loss. Four weeks later, he showed profound left hearing loss. Although he did not complain of any vestibular symptoms, horizontal nystagmus to the right was observed.

Analysis of haematological data revealed typical Disseminated intravascular coagulation (DIC) syndrome. His platelet count was 32,000. He manifested haemorrhagic diathesis and died 13 days later. An autopsy confirmed the presence of gastric adenocarcinoma with metastases to the lung and to the bone narrow.

Temporal Bone Histopathological Findings

Right ear : Blood or its precipitates were found only in limited parts of the cochlear duct. Distention and collapse of Reissner's membrane and the cochlear duct were noted. The organ of corti was lost or flattened in all turns. Strial atrophy and severe loss of spiral ganglion cells was noted. The saccule was filled with haemolysed blood.

Left Ear : Blood was found in the cochlear duct of all turns, in the scala tympani of the basal turn and in the perilymphatic spaces of the vestibule. The organ of corti was better preserved. The stria vascularis was flattened.

DIAGNOSIS OF SUDDEN DEAFNESS

Every patient suffering from sudden SNHL must be thoroughly evaluated in order to treat any underlying disorders that may be contributing to this disorder. (Cited in http://www.utmb.edu/oto/Grnds.dir/suddendeaf.htm).

Evaluation and management of sudden sensorineural hearing loss should be considered a medical urgency, if not an emergency A careful history may suggest a cause. The exact circumstance and characteristic of the onset of the hearing loss should be determined. The history should outline the complaint and include questions regarding potential precipitating events preceding the SNHL such as recent air travel, strenuous exercise, weight lifting, diving, falls, other head trauma, viral illnesses, febrile states, previous otologic surgery, medications and exposure to pesticides. Past medial history should include questions regarding diabetes mellitus, hyperlipidemia, arteriosclerosis, hypertension and syphilis. Family history of hearing loss and other diseases should also be elicited from the patient (cited in http://www.utmb.edu/oto/Grnds.dir/suddendeaf.htm).

The physical examination should be complete with special attention directed toward the otolaryngologic and neurologic portions of the examinations.

Audiometry should be performed in all patients presenting with sudden SNHL. Additional tests may be required to determine the site of lesion, such as Auditory Evoked Brainstem Response

(ABR) and Electronystagmography (ENG) especially if vertigo is present. If retrocochlear pathology, such as an acoustic neuroma is suspected, then Magnetic Resonance Imaging with Gadolinium (MRI) or less preferably Computerised tomography (CT), is indicated to examine the internal auditory canals and posterior fossa. CT imaging should be obtained for the hearing loss that may be trauma induced (cited in http://www.utmb.edu/oto/Grnds.dir/suddendeaf htm).

Laboratory studies should include a complete blood cell count, Erythrocyte sedimentation rate (ESR), coagulation profile (including prothrombin time, partial thromboplastin time, and clotting time), electrolytes with fasting blood glucose, cholesterol and triglycerides, thyroid function tests, ACTH plasma corticol stimulation test, serological tests for syphilis, and Autoimmunologic tests such as Anti Nuclear Antibody (ANA) and Rheumatoid factor (RF) (cited in http://www.utmb.edu/oto/Grnds.dir/suddendeaf htm).

Lumbar Puncture

Judgement must be exercised in the individual patient with sudden deafness in determining whether a lumbar puncture should be done. Measurement of opening and closing pressures and examination of the cerebrospinal fluid for color, clarity, cells, protein content electrophoresis and serology may provide essential information that is available by no other means. One must bear in mind that sudden deafness does occasionally occur with cerebello pontine angle tumors, meningo encephalitis and trauma.[Snow,J.B.(1973)]

According to Wetmore (1986) Diagnosis of sudden deafness is based on the following :

1) Symptoms

a) The sudden loss of hearing in one ear may be noted upon awakening or may be associated with a popping sensation in the ear followed by roaring tinnitus.

b) Vertigo may occur

c) There is usually no pain.

2) Signs

a) Nystagmus (if vertigo is present)

b) Impaired ability to discern spoken words on whisper test (two syllable words spoken into the patient's ear at varying levels o: loudness and the patient asked to repeat the words).

c) Normal tympanic membrane

d) No middle ear effusion.

3) Tests

A. Tuning Fort Tests

1. Rinne Test: Use the 256, 512 and 1024 Hz tuning forks to compare loudness between the fork placed on the skin over the mastoid cortex and the same fork held in air near the patients ear canal.

a) With a conductive hearing loss the patient perceives that the fork placed on the mastoid cortex is louder. With a mild conductive hearing loss, the patient states that only the 256 Hz fork seems louder on the mastoid cortex. With a moderate to severe conductive loss, all three forks seem louder when held on the bone.

b) With normal hearing or with a sensorineural hearing loss, the fork held in the air in front of the ear canal sounds louder.

2. Weber Test: The 512 Hz fork is struck and then placed on the patients forehead or scalp in the midline.

a) With a unilateral conductive hearing loss, the tone is louder in the ear with the conductive hearing loss (worse hearing ear).

b) With a unilateral sensorineural hearing loss, the patient hears the tone louder in the better hearing ear.

B. Audiometry

1. Most important test in documenting hearing loss.
2. Tests the patient's threshold of hearing to puretones as well as to speech.
3. Also tests the patient's ability to understand speech (discrimination score) by asking patient to repeat two syllable words presented at 40 dB above the threshold of hearing.
4. Allows the examiner to determine both the degree of hearing loss and the pattern eg. severe high frequency sensorineural loss, moderate low frequency mixed (conductive plus sensorineural) loss.

C. Electronystagmography (ENG)

1. Test to evaluate the functioning of vestibular system.
2. Performed by attaching electrodes lateral to each eye and looking for nystagmus.
3. Patient is asked to perform several tasks such as following a moving pendulum with his eyes and gazing 20 degrees right and left. He is also placed in various positions, eg. supine with head left or supine with head hanging over the end of the table. to look for positional nystagmus. Cold and warm water (or air) are then injected into each ear canal to look for a caloric response.
4. Can detect abnormalities of the vestibular portion of the inner ear.
5. May be able to localize the vestibular abnormality either in the brainstem or in the inner ear.

D Auditory Brainstem Response (ABR) Audiometry

1. This test is useful in localizing disease occurring in the VIET nerve or brainstem and can be used to detect the threshold of hearing in the higher frequencies.

2. The ear is stimulated by a tone click, and the electrical potentials of the VIII nerve and brainstem that are produced within 10 ms, of the stimulus are measured.

3. Because of low amplitude of these evoked auditory potentials, computer averaging is necessary to distinguish the signal from the background electrical noise.

E. Temporal Bone Radiographs

1. May be useful for distinguishing idiopathic sudden hearing loss from an acoustic neuroma.
2. Look for widening of the internal auditory canal as a sign of a neuroma.

F. **Computed Tomography** (CT **Scan)**

1. A contrast enhanced CT scan can detect most acoustic neuromas. If the enhanced CT scan is negative for tumor, an air contrast CT scan detects small or intercanalicular tumors.
2. It is usually not necessary in a work up of idiopathic sudden hearing loss (ISHL).

G. **Serologic Test for Syphilis**

Laurikainen et al. (1989) in their study of 80 patients with sudden deafness have highlighted the role of Electronystagmography (ENG) in the diagnosis of sudden deafness. As many as 31 of the 80 patients with acute hearing loss had abnormal ENG findings which aided in their diagnosis.

Franklin et al. (1989) in their report of sudden sensorineural hearing loss as a presentation of Multiple sclerosis comment on the role played by Brainstem Evoked Response Audiometry (BSERA) Speech audiometry, Acoustic reflex and Electronystagmography. In yielding significant objective information concerning brainstem

involvement. They highlight on the role played by Magnetic Resonance Imaging (MRI) in objectively documenting white matter demyelinisation in patients with Multiple sclerosis with sudden sensorineural hearing loss.

Harris and Sharp (1990) introduced the classical immunoblotting technique into otologic research in order to determine cross reacting circulating antibodies in patients with rapidly progressive SNHL. Some of their patients evidenced of a single or double band migrating at 68,000 molecular weight. However Moszicki et al. (1990) testing a similar group of patients found multiple band with a molecular weight between 50-60 KD.

Hall et al. (1991) assessed the red blood cell deformability (RCD) by a filtration technique in 12 patients with sudden sensorineural deafness. RCD was shown to be significantly impaired in patients with sudden sensorineural deafness with the help of this technique.

Mark et al. (1992) in their article describe 12 patients with enhancement of the cochlea and/or vestibule on Gadolinium-Diethylnetriamine pentaacetic - enhanced magnetic resonance imaging (MRI), correlating the enhancement with the auditory and vestibular function. According to them, while abnormalities on Electronystagmography (ENG) and audiograms are non-specific and only indicate a sensorineural problem, enhanced MRI may separate patients with retrocochlear lesions such as acoustic neuroma's from those in whom the abnormal process is in the labyrinth or the brain.

Labyrinthine enhancement seen in the case reported represents accumulation of Gadolinium in the labyrinth following leakage through the abnormal labyrinthine membrane. Thus, Gadolinium enhancement of the labyrinth appears to be a highly specific sign of! labyrinthine disease.

Nakashima et al. (1992) believe that the laser-doppler measurement of cochlear blood flow will elucidate the status of cochlear blood flow in various inner ear diseases including sudden deafness.

Veldman et al. (1993) in their study of 76 patients with al clinical diagnosis of idiopathic rapidly progressive SNHL (n=15) and sudden deafness (n=31) and with other etiologies of their hearing loss (n=30) made use of the "Immunoblotting technique" to detect antigenic epitopes in these patients and noticed that the antigenic epitopes detected with immunoblotting were not cochlea specific but they were also found in protein extracts of other organs (cranial nerves, kidney, brain etc.).

Fabiani et al. (1993) reports of the applicability of Evoked Otoacoustic Emission (EOAE) in the clinical field for studying adult sensorineural hearing loss. They suggest that EOAEs should be applied as a routine audiological test for adult sensorineural hearing loss not only in cases with a well defined diagnostic profile, but also in those cases with a less clear diagnostic pattern, in order to collect more data and possibly gain better knowledge about the pathophysiology of the inner ear. In relation to sudden deafness the

authors comment that up to now, EOAEs have not been shown to be useful in establishing the etiology of sudden deafness as they cannot usually be measured owing to the degree of hearing loss But recordable EOAEs have favourable prognostic value if detected again within two weeks of the onset of deafness (Takeda et al. 1992).

Ohinata et al. (1994) highlight the importance of measuring blood viscosity and plasma viscosity in patient's with sudden deafhess as a positive correlation between average hearing level in puretone audiogram and blood viscosity or plasma viscosity was positive in their study of 51 patients with sudden deafness. Patients with sudden deafness were found to have increased blood viscosity/plasma viscosity thus suggesting the importance of viscosimetry in the diagnosis of sudden deafness.

Drulovic et al. (1994) report of two patients with the diagnosis of clinically definite Multiple sclerosis, in whom unilateral sudden deafness was the first and only manifestation of the disease. They describe "multiple sclerosis as the cause of sudden pontine deafness" based on the results obtained from evaluation by means of brainstem evoked response audiometry (BSERA). Magnetic resonance imaging and tonal audiometry, thus highlight their role in the diagnosis of sudden deafness.

Marangos (1996) states that in some cases, Electrocochleography (ECoG) may be the only clinical tool indicating the site of lesion for a sudden hearing loss, because it enables the differentiation between sensory and neural hearing-impairment and

transtympanic ECoG provides valuable information not only in cases of sensory hearing loss or hydrops, but also in retrocochlear lesions such as those associated with multiple sclerosis or acoustic neuromas.

Luxford and Saunders (1996) in their review of patients with sudden deafness from 1989 to 1993 comment that positive antinuclear antibody titer is likely to be seen in patients with sudden deafness, thus indicating that antinuclear antibody titer may sudden the diagnosis of sudden deafness.

Hughes et al. (1996) summarise a list of diagnostic tests and their clinical indications for sudden deafness. The list is as follows :

	Indications	**Tests**
1.	All patients	Basic audiometry
2.	Acoustic neuroma or skull base lesion	MRI with contrast CT with contrast (to evaluate for both tumor and bony anomaly in pediatric population).
3.	Immune inner ear disease (LTT)	Lymphocyte transformation test Western blot immunoassay. Antigen non-specific serologic tests. Acute phase reactants.
4.	Syphilis	Fluorescent Treponemal Antibody absorption (FTA- abs). Microhemagglutination ass ay - Treponema pallidum (MHA.-TP).
5.	Other bacterial infections	Lyme titer, Cultures as ind|cated clinically .Acute and convalescent titers.
	Virus	AIDS testing as indicated clinically.

Indications	Tests
6. Various indications	Brainstem Auditory Evoked Potentials (AEPs). Electronystagmography (ENG), particularly in ototoxicity Electrocochleography (ECoG) in Meniere's disease. Perilymph fistula "tests", complete blood count, blood chemistries, metabolic studies.

Getson et al. (1997) in their review of 179 patients who underwent surgical repair for a presumed unilateral Perilymphatic fistula state that the ENG fistula test, the Platform Fistula Test (PFT) and Electrocochleography (ECochG) are the diagnostic tests available apart from the audiometric examination to reveal the presence of a perilymphatic fistula which may lead to sudden deafness.

Kou and MacDonald (1998) retrospectively studied cases at the hospital for sick children in toronto between the years 1980 and 1995 and comment that high resolution CT scans of the temporal bones with bone algorithms and coronal axial view were performed on all children with sudden hearing loss after 1988 to aid in the diagnosis of sudden deafiiess in these children.

Takasaki et al. (1998) highlight the importance of IgG antibody titres to Human Herpes Virus 6 (HHV-6) and 7 (HHV-7) in patients with Idiopathic facial nerve palsy or sudden deafness and suggest that IgG antibody titres may play a significant role in the diagnosis of sudden deafness/facial nerve palsy.

Franklin et al. (1989) report of two patients with multiple sclerosis with sudden sensorineural hearing loss.

The following is a brief summary of their findings :

Patient 1	Patient 2
1. Sudden sensorineural hearing loss was the sole manifestation of multiple sclerosis	Multiple sclerosis was diagnosed eight years prior to the onset of sudden right sensorineural hearing loss.
2. Audiogram obtained one week after onset revealed moderate right sensorineural hearing loss with puretone average of 55 dB.	Audiometry revealed mild to moderate right sensori-neural hearing loss with puretone average of 40dB.
3. Right crossed acoustic reflexes were absent.	Absence of right acoustic reflex.
4. Poor speech discrimination to phonemically balanced words and synthetic sentence identification.	Poor speech discrimination to phonemically balanced words & synthetic sentence identification.
5. Auditory brainstem responses obtained two months after onset, had left prolonged absolute latencies of Waves I, III, V as well as prolonged interpeak interval on right.	Auditory brainstem response indicated signifi-cantiy prolonged latency of wave V with no identi -fiable wave I or HI.
6. Magnetic resonance imaging was diffusely abnormal with multiple foci of prolonged spin density and T2 signal in the white matter both preventricularly and subcortically.	Magnetic Resonance Imaging showed extensive involvement of Central Nervous system with Multisclerotic plaques.
7. Electronystagmography revealed decreased rotatory responses, positional nystagmus & impaired optokinetic nystagmus.	Electronystagmograpbic responses were asymmetric and bilaterally decreased.

Serological tests including antinucleus antibody and rheumatoid factor were negative in both the patients.

Schorn and Zwicker (1990) in their study of 25 patients with unilateral sudden deafness report that frequency resolution was affected at 500 Hz and 4 kHz in these patients and in the presence of a background noise there was a further deterioration is frequency resolution (i.e.) the tuning curve became flatter.

The temporal resolution in these subjects was only slightly reduced and with background noise the temporal resolution is also affected in the low frequency range. The temporal resolution in background noise condition was noticed to be twice as poor at 500 Hz and three times as poor at 4 kHz as normal hearing subjects.

Combined frequency/temporal resolution factors with background noise showed that regardless of the degree of hearing loss, all of the patients with sudden deafness exhibited some impairment in their frequency and/or temporal resolution at both the tested frequencies (500 Hz and 4 kHz) while in other syndromes (like toxic inner ear damage, noise induced hearing loss, presbycusis, degenerative progressive inner ear hearing loss etc.) the impairment was especially manifest in the 4000 Hz frequency range.

Shiraishi et al. (1993) in their report of 98 patients with sudden deafness observed that the initial hearing levels (arithmetic mean of 200 Hz, 500 Hz, 1 kHz, 2 kHz and 4 kHz) ranged from 25 to 111 dB HL with mean of 68.3 dB.

Moffat et al. (1991) in their study of 284 patients with vestibular schwannoma stated that 12% of these patients presented

with sudden deafness. The most common finding was a high frequency loss found in 35.2%, of the sudden deaf group and 42.4% in the non-sudden deafness group. A flat trace was seen in 26.5% of the sudden deafness group and 24.8% of the non-sudden-deaf group. Low frequency, cup shaped and corner audiograms were rare in both the groups. Dead ears were more common in the sudden deafness group (29.5%) than in the non-sudden deafness group.

Drulovic et al. (1994) *give* a report of the following studies done on two patients who presented with sudden deafness and tinnitus as the sole initial symptoms.

Patient 1	**Patient 2**
The tonal audiogram indicated a hearing loss in left ear with a horizontal curve between 50 & 60dBHL. Right ear audiogram indicated normal hearing levels.	The tonal audiogram indicated normal hearing in left ear and severe hearing loss with hearing between 60, 30 & 90 dB HL at 8 kHz. At 4 kHz, the hearing threshold was 40dB HL in right ear.
Tympanogram was bilaterally normal.	Tympanogram was bilaterally normal.
Reflexes were ipsilaterally absent on the left side.	Stapedial reflexes were absent on right side with ipsilateral stimulation & appeared with normal threshold at 500 Hz, 1 kHz, and 2 kHz on the left side when the stimulus was ipsilateral.

Patient 1	Patient 2
Brainstem responses: When left ear was stimulated, only first three waves were present with normal latencies; while IV and V waves were absent. When right ear was stimulated to 100 dB nHL clicks brainstem responses appeared normally.	**Brainstem responses** : Evoked by 100 dB nHL click stimuli applied to right ear showed only wave I with norma latency. Same procedure applied to left ear showed normal responses.
Electronystagmography : Revealed no nystagmus.	**Electronystagmography** : Revealed no nystagmus.
Rotatory test revealed the function of vestibular apparatus to be bilaterally normal.	**Rotatory test** revealed the normal functioning of the vestibular appartus.

Luxford and Saunders (1996) in their review of 823 patients with sudden sensorineural hearing loss from 1989 to 1993 repor that the audiograms of patients with sudden bilateral sensorineural hearing loss was usually asymmetric in configuration.

Marangos (1996) report of two patients with neurologica] symptoms presenting with sudden deafness.

The following is a brief summary of their report.

Case 1	Case2
39 year old male with diagnosed definite multiple sclerosis suffered from fluctuant neurological symptoms including ataxia and diplopia.	55 year old male hospitalized due to definite neurological symptoms probably due to chronic alcoholism. Two days following admission, he developed acute unilateral hearing loss.

Case 1	Case 2
Elevation of PTA of 60 dB rapid tone decay. Negative recruitment and poor speech discrimination all pointed to a neural lesion in this case.	Puretone audiometry revealed a ' dead ear' and ' sensory' hearing loss on left stapedial reflexes.
Brainstem auditory evoked potentials revealed normal thresholds & interpeak latencies on left, but no responsesat all on right. Even wave I was absent in the affected ear,	Reliable Brainstem Auditory Evoked Potential responses could not be obtained.
Reception potentials (cochlear microphonics, summating potentials) appeared to be normal. These indicated a lesion in the hearing nerve itself.	In Elecrocochleography the receptor potentials (Cochlear Microphonics, Summating potentials) were normal.

Cox and Sargent (1997) in their report of three patients with sudden sensorineural hearing loss following non-otologic surgery in which cardiopulmonary bypass surgery was not involved report the following symptoms seen in the three cases :

Case 1	Case 2	Case 3
This case had a complaint of bilateral hearing loss following reversion of a nasal fracture under general anasthesia, complaint of aural fullness and light headness.	This case had a complaint of fullness & hearing loss that began immediately after an otherwise uncomplicated lumbar spine surgery using general anaesthesia two weeks before presentation. No complaint of vertigo reported.	This patient presented three weeks after undergoing uncomplicated decomposition of the lumbar spine for spinal stenosis using general anaesthesia. Patient reported awakening from surgery with muffled hearing.

Case 1	Case 2	Case 3
Physical examination was normal.	Physical examination was normal.	Physical examn. was normal.
Puretone average was 39 dB in right ear & 44 dB in left ear.	Audiogram showed severe sensorineural hearing loss in right ear with normal hearing in left ear.	In right ear the patient's puretone average was 104 dB.
The Speech Reception Threshold was 40 dB bilaterally & speech discrimination scores were 92% and 80% in the right and left ears respectively.		His speech reception threshold was 95dB & speech discrimination score was 0%.
Magnetic Response Imaging was Normal	Magnetic Resonance Imaging & Computed tomography was normal.	
Audiometric Brainstem Responses were consistent with behavioural thresholds, lymphocyte transformation test for anti cochlear antibodies were normal except for antinuclear antibody titre.		
After treatment with prednisone for one month only slight improvement in hearing was noticed.		
Speech reception threshold was 30 dB bilaterally. Puretoneaverage was 34 dB in right ear and 36 dB in left ear. Speech discrimination scores were 100% bilaterally.		

A summary of the above cases is presented in order to have a better understanding of sudden deafness and various studies related to it.

PROGNOSIS

It is difficult to accurately predict the outcome of sudden SNHL given the low incidence and unpredictability of its natural course. Also, there are problems evaluating recovery statistics especially for treatment protocols, because of the significant percentage of patients who recover spontaneously (65%) (cited in http://www.utmb.edu/oto/Grands.dir/sudden deaf.htm).

Approximately one third of patients have a return of hearing to normal, one third are left with moderate hearing loss and one third have total loss of useful hearing (cited in http://www.med students. com. br/otor/otor4. htm).

Once recovery of hearing begins, it is likely to take place very rapidly in a matter of few days. The longer the delay between the onset of deafness and the onset of recovery, the worse is the prognosis for complete recovery. Nevertheless, complete recovery of hearing does occur at times even after several weeks of profound loss. Spontaneous recovery to normal hearing is more likely to occur if the deafness is not total initially. The prognosis cannot be easily related to the rapidity of onset, the presence of tinnitus, recruitment or the type of Bekesy audiogram.

Laurikainen et al. (1989) in their study of 80 patients treated for sudden deafness have reported that patients with an abnormal ENG (electronystagmography) study showed a poor recovery of speech reception threshold, whereas those with a normal ENG study showed slightly significant recovery.

Shivaishi et al. (1991) compared the time courses of hearing recovery resulting from two types of treatment of sudden deafness patients - Defibrinogenation (DF) therapy and Betamethazone (BM) therapy and their study indicated that DF therapy showed a tendency to better hearing recovery, when the absolute value of hearing level (0.25-4 kHz) was used for comparison of hearing recovery in the two groups.

Shiraishi et al. (1993) studied the prognostic factors in 98 patients with idiopathic sudden deafness and concluded that the interval between the onset of hearing loss and start of treatment, initial hearing level, and the existence of vertigo all had a significant correlation with the degree of hearing recovery. They suggested thai there may be a high rate of spontaneous recovery within the first days after the onset of hearing loss. Their study confirmed the opinions of other investigators that patients seen and treated early in the course of their illness enjoy a significantly better prognosis than those seen later.

Though it has been suggested that decreased hearing in the opposite ear may indicate fragility of the bilateral inner ears and poor recovery potential, the study conducted by Shairaishi et al. (1993) showed only a weak correlation between the pure tone average of the opposite ear and the improvement rate.

Their study confirms the statement that sudden deafness accompanied by vertigo or dizziness has a poorer prognosis than without these symptoms. They also report of higher improvement rates for upward slope and flat types of hearing losses in their study.

Nakashima and Yanagita (1993) in order to elucidite the clinical significance of vertigo in sudden deafness, investigated the relationship between vertigo and hearing loss in sudden denfness. The outcome of unilateral sudden deafness with and without vertigo was evaluated according to the severity of the initial hearing loss, the shape of the audiogram and other variables. Their study indicated that hearing recovery of high tone frequencies was worse in patients with vertigo than in those without vertigo even when the initial h earing loss was the same.

Once the recovery of hearing begins, it is likely to take place very rapidly in a matter of few days. Even when severe initially, the vertigo tends to subside within one week and as a general rule, all vestibular symptoms clear spontaneously within 6 weeks. (Cited in http://www.medsrudents.com.br/otor/otor4.htm).

According to Byl et al. (1984) hearing recovery in idiopathic sudden deafness is worse in patients with vertigo than in those without vertigo.

Ohinata et al. (1994) investigated the relationship between average hearing level in puretone audiogram and blood viscosity or plasma viscosity in patients with sudden deafness. Their study indicated that when the distribution of the average hearing level was 40 to 74 dB, a few of the patients with "recovery" or "good improvement" and most of the patients with "fair improvement " or "no change" belonged to the low viscosity group. And most of the patients with flat type hearing-impairment and a few patients with

high tone type hearing-impairment belonged to the high viscosity group. These results suggest that prognosis is better in patients with sudden deafness who have a low blood/plasma viscosity compared to sudden deafness patients with high blood/plasma viscosity. Elevated erythrocyte sedimentation rate (>25) also signals a poorer prognosis (cited in http:/www. utmb.edu/oto/arnds.dir/ suddendeaf.htm).

Sano et al. (1996) divided a total of 547 patients with idiopathic sudden hearing loss into eight groups according to the days to the initial visit (1-8 days) after onset and investigated initial hearing levels, hearing recovery and its process among these groups. Their study indicated no significant differences in initial hearing thresholds and hearing recovery among the eight groups. However, a difference was noticeable in the distribution of the thresholds at the initial visit and that on the 7th day between groups, 1, 2 and 7. Many cases showed rapid recovery in groups 1,2 and 7. They speculated that the treatment influenced the hearing recovery process inpatients who showed relatively rapid recovery.

Hirayama, et al. (1996) in their study of 127 patients with idiopathic bilateral sensorineural hearing loss stated that hearing improvement was achieved in some cases with rapidly progressed idiopathic bilateral sensorineural hearing loss, when the treatment had started in the early stages after the onset of rapidly progressive hearing loss.

Harada (1996) examined the patterns of hearing recovery in idiopathic sudden sensorineural hearing loss (ISSHL) during the initial stage of treatment in 51 patients who showed significant recovery. This study suggested that cases of treated ISSNHL in which recovery of hearing takes place rapidly at the initial stage tend to have a favourable outcome. Initial rapid recovery was observed more frequently in cases who showed less hearing loss at first examination greater degree of hearing-impairment or less hearing at the fixed stage. The pattern of hearing recovery at the initial stage of ISSNHL appeared to reflect the prognosis to a certain degree.

Kallinen et al. (1997) in their study of 168 consecutive patients with sudden deafness found that the configuration of the audiogram of sudden deafness patients is prognostic of the outcome and that patients with low frequency sloping hearing-impairment have a better prognosis compared to patients with a high sloping loss. The treatment modality along with the configuration of hearing loss appeared to effect the prognosiSjfor anticoagulant treatment was most effective in low sloping hearing loss and showed good prognosis, wh ile carbogen inhalation was more effective for patients with high sloping losses and showed good prognosis for such patients.

Stokroos et al. (1998) in their study of 44 idiopathic sudden sensorineural hearing loss (ISSHL) patients noted that a majority of their patients suffered tinnitus "which has a poor prognosis. They compared the prognosis of patients treated with "Aciclovir" and "Prednisolone". In their study, a majority of ISSHL patients treated with "Prednisolone" experienced some subjective hearing recovery

and most did so after one week of treatment. "Aciclovir" treated patients did not experience hearing recovery earlier or more often. Application of "Aciclovir" did not change the tinnitus prognosis. Patients with frequent concomitant complaints of pressure sensation on the affected ear and a sens of disequilibrium or vertigo had a favourable prognosis, irrespective of "Aciclovir" use. Patients treated with "Prednisolone" achieved better hearing recovery compared to untreated ISSHL patients reported previously. In severe/profound hearing loss, the chances of hearing recovery was poor and in cases of mild/moderate hearing loss the chances were better.

Thus, the most important prognostic indicators for sudden deafness include severity of initial hearing loss, severity of vertigo and time from onset to the start of treatment when the initial audiogram is usually obtained. Other indicators such as age, configuration of initial audiogram and vestibular testing results are also important factors influencing the prognosis.

SYNDROMES ASSOCIATED WITH SUDDEN SENSORINEURAL HEARING LOSS

The following syndromes have been found to be associated with sudden sensorineural hearing loss.

1. **Cogan's Syndrome**

The syndrome is primarily a non-syphilitic keratitis with vestibulo auditory symptoms (Dorland, 1994).

2. **Mondini Syndrome**

A characteristic of Mondini malformation of the inner ear. It can be sudden hearing loss. A mondini malformation is an underdeveloped cochlea. With a Mondini, hearing loss can be sudden or gradual, mild or profound. Mondini malformation can be detected with a CAT scan. It is thought to be 95% congenital, possibly x-linked. An interesting characteristic which sometimes manifests itself with mondini malformation is that some people hear ultra high frequencies only (i.e. over 8000 Hz). This may be due to the high frequency hair cells being located at the fat part (beginning) of the cochlea and since the cochlea does not develop completely, this area is saturated with hair cells (Cited in http://www.weizmann.ac.il/deaf-info/sudden deahess.html).

TREATMENT OF SUDDEN DEAFNESS

The lack of scientific understanding of the etiology or etiologies of the problem of sudden deafness limits its treatment to simple empiricism. Every proposed treatment has studies to support it, but corroborative data from independent researches is usually lacking. Perhaps the fatal flaw in most of those studies is that a single treatment is applied to a symptom that undoubtedly has multiple etiologies (cited in http:www.urnrnah.net/software/software/softbase/sudden.txt).

The frequent spontaneous recovery of hearing to normal or near normal levels makes evaluation of any form of therapy for sudden deafness very difficult. Each form of therapy appears to be effective in a large number of patients, and no form of therapy has been found to be effective in all patients with sudden deafness. In fact, it is difficult to j udge whether any form of therapy advocated for sudden deafness produces a higher recovery rate than would have occurred spontaneously. Many treatments have been advocated for increasing the blood flow to the inner ear and these include blood thinners and drugs designed to dilate blood vessels such as Niacin, Carbogen gas, Histamine, Bellargal and many others. Despite numerous studies, none of these drugs have been shown to be useful in the treatment of sudden sensorineural hearing loss and therefore, these are usually not advocated for the treatment of this disorder.

(Snow, J.B 1973)

The therapy currently advocated includes Vasodilation, Anticoagulation, Reduction of the viscosity of the blood, Sedation

and Tranquilisation, Vitamins, Corticosteroids, and Bed rest. Vasodilation, Anticoagulation, Reduction of the viscosity of blood are based on the theory of vascular causes for sudden deafness. The rationale for Vitamin therapy and Cortico steroid therapy is less clear.

Vasodilation has been advocated by Van Dishoeck and Bierman (1957), Sheehy (1960), Jaffe (1967) and Rubin (1968). Van Dishoeck and Bierman mention spasmolytica but do not specify the agents. Sheehy advocates Intravenous-Histamine-Phosphate initially, to be followed by Intramuscular Histamine Phosphate. Subsequent Oral therapy consists of "sublingual Histamine Phosphate" and Nicotinic acid. Sheehy reported that on this treatment regimen, 24 of 70 patients or 34 percent recovered. Jaffe also advocates "Intravenous Histamine" and "Nicotinic acid". Rubin advocates "Hyoscine" or "Atropine" intramuscularly or intravenously in the acute phase and Procaine Hydrochloride intravenously after several days or weeks. He also recommends Nylidrin, a "Non Catecholamine" a n) β (B e e t a) receptor stimulant. Rubin reports t o normal hearing i n 50% of his type I patients, but no appreciable effects in type II and III (snow J.B. (1973)).

The results of vasodilation therapy appears to approximate spontaneous recovery rates.

Study of the effect on cochlear blood flow in guinea pigs of adrenergic agents, adrenergic blocking and antiadrenergic agents, cholinergic agents and anticholinesterase and cholenolytic agents indicates that the adrenergic and cholinergic nervous systems have

relatively weak control over cochlear vessels which is also true of their effect on cerebral vessels (Suga and Snow, 1969).

In their study of vasodilating drugs and some related agents, Suga and Snow showed that Nicotinic acid, even in massive doses has no measurable effect on cochlear blood flow. Histamine-phosphate and Betahistine increases cochlear blood flow in dosages that produce broncheospasm in the guinea pig and may well produce vasodilation on the basis of anoxia

The rationale for vasodilation therapy for sudden deafness is questionable in view of the preponderance of evidence for the viral etiology of most cases of sudden deafness. Some, now advocate vasodilation in view of the vascular changes that occur in viral diseases. Granting these changes in individual capillaries, overall blood flow may not be decreased but may actually increase in the area of inflammation. Should cochlear vasodilation be achieved clinically by either "Histamine phosphate" or "Betahistine", there is serious question whether such an effect is desirable in the presence of micropetechiae in viral infections. Resolution of the question of the efficacy of this therapy must await well controlled clinical studies.

Bolognesi (1960) advocated "Anticoagulant therapy "after improvement in the hearing of three of five patients on Heparin and Coumadin therapy. Otherwise, advocacy of anticoagulation therapy has largely remained unpublished. Viral infections often produce temporary bleeding tendencies. In view of probable viral etiology of most sudden deafness, such therapy may well be contraindicated.

Jaffe (1967) advocates low molecular weight "Dextran" to reduce the viscosity of the blood. Van Dischoeck and Bierman (1957) advocate "Corticosteroid therapy" for sudden deafness.

Vitamins in non-toxic doses do not harm.

Laurikainen et al. (1989) conducted a study on eighty patients treated for sudden deafness over a period of 5-7 years. All of them had received "Anticoagulants (Intravenous Heparin 2 ml, 4 times daily for 2 days and then oral "Warfarin" according to thromboplastin time) and oral "Betahistine hydrochloride", 8 mg, 3 times a day for 30-90 days. They suggest that due to lack of effective antiviral drugs and the difficulty of confirming viral etiology, viral infection should be ignored in choosing treatment for sudden deafness. Their study indicates that the benefit of anticoagulant treatment is always limited, even in the pure cochlear form of sudden deafness. Because anticoagulative treatment continued for several months carries life threatening risks, safer alternatives should be considered for the improvement of cochlear circulation during vasospasm, thrombosis or other metabolic disorders.

Myer et al. (1989) advocate that in children with sudden, progressive or fluctuating sensorineural hearing loss and multiple sensory deficits including blindess or contralateral sensorineural hearing loss or prior head trauma prompt "Surgical Exploration" is mandatory. Additionally, the aggressive management of otitis media with effusion is essential in such patients to minimise fluctuations in hearing caused by superimposed conductive hearing loss.

Shiraishi et al. (1991) performed a paired double blind comparative study on 168 patients with sudden deafness. They compared the recovery of these patients treated with "Defibrinogenation and Steroid Therapies". In Defibrinogenation therapy a medicine called "Batroxobin" which is a thrombin like enzyme isolated from snake venom and which enhances blood circulation by decreasing serum fibrinogen is administered. The steroid administered to these patients with sudden deafness was "Betamethazone". The average hearing level was 79.2 dB HL in the Defibrinogenation (DR) therapy group and 82.3 dB HL in the Betamethazone (BM) therapy group. The DF group showed significantly better hearing one week after start of treatment at frequencies of 0.25 kHz, .5 kHz and 1 kHz. After two weeks of treatment, the average hearing level for the frequency range of 0.25-4 kHz was 47.0 dB HL with DF therapy and 55.8 dB HL with BM therapy. Thus the DF group still showed better hearing than the BM group but not as significantly as after one week. They concluded that therapy for increasing blood circulation is important in the early phase of sudden deafness, especially for hearing loss in the low to middle frequency range.

Erikson et al. (1992) present the first case report of recovery of a patient from repeated sudden hearing loss with corticosteroid use in the presence of an acoustic neuroma. The case report is of a patient with pre-existing deafness in one ear. The patient developed hearing loss in the only hearing ear that resolved with oral corticosteroids. Magnetic Resonance Imaging (MRI) was done after the third episode of sudden hearing loss revealing a small Acoustic-

neuroma. After surgical decompression, a similar episode of sudden hearing loss occurred, with resolution after another course of steroids. They, therefore, suggest that the clinician must have a high index of suspicion for Acoustic neuroma even if hearing returns to normal.

Shiraishi et al. (1993) conducted a study on 98 patients with idiopathic sudden deafness treated with modified Defibrinogenation (DF) therapy including Batroxobin, Low Molecular Dextran, Vasodilation and Vitamins. They concluded that modified DF therapy is one of the most effective regimens for sudden deafness. One distinctive feature of this therapy, according to them, is the effectiveness for both mild and severe hearing loss with flat type audiogram. However, patients with initial PTA values of more than 90 dB showed poorer recovery even with this therapy. They suggest that attempts should therefore be made to find an effective treatment modality for profoundly deaf patients.

Ravi and Henderson (1996) in their report of a case with sudden deafness as the sole presenting symptom of Diabetes mellitus report that "Insulin therapy" resulted in restoration of hearing in this case. Since hyperglycemia is the main cause for reversible or irreversible complication, they presume that Insulin therapy played a major role in recovery of this patient's hearing.

Hughes et al. (1996) present a partial list of treatments for sudden sensorineural hearing loss, based on the review of the literature of many studies. The following is the list of treatments presented by them :

1. Antiinflammatory/ Immunologic agents	Cortisone,Prostaglandin
2. Diuretics	Hydrochlorothiazide/Triamterene Lasix
3. Antiviral agents	Acyclovir
4. Vasodilators	5% carbondioxide and 95% oxygen (Carbogen), Papaverine, Buphenine (Nylidrin), Naftidrofiiryl (Nafronyl), Thymoxamine, Prostacyclin, Nicotinic acid, Pentoxyfylline.
5. Volume expanders/ hemodilators	Hydromyethyl starch, Low molecular weight Dextran.
6. Defibrinogenators	Batroxobin
7. Calcium antagonists	Nifedipine
8. Other agents and procedures	Amidotrizoate, Acupuncture, Iron, Vitamine, Procaine

Many treatment regimens have been proposed for idiopathic sudden hearing loss as indicated above in the table, but at present, according to the authors, independent studies have not consistently supported the use of any one treatment modality. Wilkins and Associates (1987) treated 109 patients with idiopathic sudden hearing loss with a "shotgun" regimen consisting of Dextran, Histamine, Hypaque (Diatrizoate - Meglumine), Diuretics, Steroids, Vasodilators and Carbogen inhalation. 33 patients received the entire protocol, and 76 received most, but not all, of the protocol drugs. Results suggested that "shotgun" therapy was no better than spontaneous recovery as reported in the literature. In a prospective, randomized, double blind study, Probst and Colleagues (1992) found no difference in hearing gains between treatment with Saline infusion together with Dextran-40, Pentoxifylline or both.

In a prospective double blind trial, Kronenberg and associates (1992) found no difference in results between a group treated with Intravenous Procaine and low molecular weight Dextran and a Placebo control group. In an eight year prospective study of 225 patients, Byl (1984) found no evidence that treatment of any kind achieved a better result than spontaneous recovery.

Despite these studies, the authors (Hughes et al. 1996) believe that three treatment modalities are justifiable and should be used in selected patients. Steroids, Low salt diet or Diuretics and Carbogen (5% carbondioxide and 95% oxygen). In addition both of the senior authors (Hughes and Freedman 1996)recently have begun routinely prescribing "Acyclovir" for idiopathic sudden hearing loss.

The specific action of steroids is unknown but they may help infections, inflammatory and immune mediated conditions. Steroids are relatively contraindicated inpatients with pregnancy, Peptic ulcer disease, Glaucoma, Diabetes, Hypertension, Inactive Tuberculosis, Osteoporosis or recent vaccination; however, prophylactic medications and careful monitoring of coexistent disease usually permit treatment for the full course. Patients also should be informed that aseptic necrosis of the femoral head is a very rare, idiosyncratic reaction to the use of steroids that requires total hip replacement for rehabilitation.

Despite disappointing results of steroid treatment in early studies which used lower doses, at present, randomised, controlled study of high dose, long term (30 days) steroids, abundant clinical

experience (Hughes et al. 1993; Moscicki et al. 1994 and Veldman et al. 1992), now supports giving Prednisone in mg/kg/day, for 1 month to patients with immune mediated SHL and for atleast 10 days for idiopathic sudden hearing loss (SHL). At present the senior author (Hughes) prefers to treat idiopathic SHL with Prednisone, 1 mgkg/ day for 30 days if hearing loss is bilateral or for atleast 10 days if the hearing loss is unilateral.

The authors frequently prescribe low salt diet - Duiretic therapy for patients with SHL because it may help and is convenient and inexpensive.

Regarding carbogen, three factors influence perilymphatic (inner ear) oxygenation : Arterial - oxygen partial pressure, Arterial carbondioxide partial-pressure, and systemic blood pressure. The carbondioxide concentration in the blood appears to be the strongest stimulus for cochlear blood flow.

In 1983, Fisch demonstrated a four fold increase in perilymphatic oxygenation with 5% carbondioxide 95% oxygen carbogen) compared with 100% oxygen. Adding oxygen rather than room air to 5% carbondioxide minimized the hypertension and acidosis induced by Carbondioxide inhalation. Fisch (1983) then compared intravenous vasodilation (Papaverine and low molecular weight Dextran) with Carbogen inhalation in 46 consecutive patients with sudden hearing loss. Each patient was hospitalised for 5 days. Carbogen was given eight times daily at intervals of 1 hour. The pure tone average improvements at 5 days were 22.3 dB for the

carbogen group and 14.5 dB for the vasodilator group, which were not significantly different. These improvements had been attributed to those that would have spontaneously occurred. At one year, however, the carbogen group had significantly better recovery (30 dB) than the vasodilator group (16.6. dB). The study also concluded that carbogen inhalation did not reduce oxygen supply to the inner ear (stealing effect) (Fisch, 1983).

In selected patients the authors (Hughes, Freedman and Haberkamp, 1996) order Carbogen inhalation therapy usually for 10 minutes, six times daily, over 3 days. Unfortunately, carbogen should be administered in a hospital setting which is inconvenient and expensive. Although oral outpatient steroid and Diuretic treatments are readily accepted by most patients, inpatient carbogen therapy is more controversial and requires the patient's informed consent.

Finally, Hughes often prescribes Acyclovir, 1 to 2g orally daily in five divided doses, because some cases of SHL are caused by hepesvirus.

In a prospective, placebo controlled, randomized study of "id opathic" acute facial (Bell's) palsy - some cases of which are caused by herpes virus - Adour (Personal Communication, 1994) found significantly improved recovery with Prednisolone-Acyclovir therapy than with Prednisone - placebo therapy. "Idiopathic" SHL may be analogous to "idiopathic" Bell's palsy, in that some cases probably are caused by herpes virus. Risks of Acyclovir are minimal and benefits are possible.

The most practical management rationale how Hughes would evaluate and manage himself for unilateral idiopathic SHL is:

1. Evaluation by basic audiometry and enhanced MR imaging.
2. Low salt (2g/day) diet and "Hydrochlorothiazide" 25 mg - Triameterene, one tablet daily.
3. Prednisone 1 mg/kg/day; and
4. Acyclovir 1-2 g orally daily in five divided doses, for atleast 10 days.

Also, inhaling carbogen periodically over several days is recommended.

Treatment of idopathic SHL should be started as soon as possible, should be aggressive and should cover those disorders that are most likely to benefit. Treatment probably is not helpful after 30 days have passed, because active disease may have resolved and damage may be permanent. Low salt diet-Diuretic and steroid treatments are recommended even after one month, however, for persistent endolymphatic hydrops (fluctuating hearing, aural pressure) immune mediated disease and progressive hearing loss. In addition, carbogen over 3 days is recommended if idiopathic SHL occurs in an only hearing ear or if the patient is highly motivated to be treated aggressively (for example, a professional musician).

Luxford and Saunders (1996) reviewed the medical records from 1989 to 1993 and stated that among 823 patients with sudden sensorineural hearing loss 14 (1.7%) had sudden bilateral

sensorineural hearing loss. They state that most bilateral cases received Steroid and Vasodilator treatment while unilateral cases were likely to receive only one of these treatments. 67% of bilateral sudden hearing loss cases improved while the improvement rate in unilateral case was 52%, however this difference was not statistically significant

Sano et al. (1996) investigated retrospectively 547 patients who visited their clinic within 8 days after onset during the period from 1972 to 1990. Most cases were treated with Steroid, Vasodilator, Metabolic activator and vitamin B from the date of the initial visit. They speculated that treatment influenced the hearing recovery process in patients who showed relatively rapid recovery.

Hirayama et al. (1996) investigated a series of 127 patients with idiopathic bilateral sensorineural hearing loss (IBSH) from 1963 to 1990 who had been treated with the same therapeutic regimen used in sudden deafness. Most of them had been treated with Steroids, Vasodilator, Metabolic activator, Vitamin B and Hyperbaric Oxygenation. They stated that improvement of hearing loss after treatment was generally less effective in the cases with rapidly progressed IBSH than in cases with SD. Hearing improvement was achieved in some cases with rapidly progressed IBSH when the treatment had started in the early stages after the onset of rapidly progressive hearing loss.

Rahko and Kotti (1997) compared Carbogen inhalation and Intravenous Heparin Infusion therapies on sudden sensorineural hearing loss. 44 patients received the former and 43 the latter treatment respectively.

The PTA (0.5 - 2.0 KHz) at the pretreatment stage was 62 dB in the heparin and 55 dB in the carbogen group. One month later, the corresponding figures were 34 and 32 dB, the difference being statistically not significant. Thus neither of these methods proved superior in the treatment of sudden SN hearing loss.

Kallinen et al. (1997) designed a study to compare a group of 168 patients with sudden deafness, the effect of Anticoagulant therapy and carbogen inhalation therapy. They found Anticoagulant Treatment was most effective in low sloping hearing loss while Carbogen inhalation to be more effective for patients with high sloping losses.

Stokroos et al. (1998) conducted a study designed prospectively, randomised, double blind and placebo controlled to test the therapeutic value of the Antiherpetic drug Aciclovir (Zovirax) on hearing recovery in 44 idiopathic sudden sensorineural hearing (ISSHL) loss receiving prednisolone. Results indicated no beneficial effect in combining Aciclovir with Prednisolone in ISSHL.

Schneider et al. (1998) conducted a randomised, blinded study in guinea pigs to predict the value of Methyl-prednisokine in the treatment of an experimental sensorineural hearing loss following drill induced ossicular chain injury. Methyl prednisolone showed no protective effect in reducing or improving the auditory threshold shifts, which occurred within seconds after drilling and remained stable throughout the 5 week observation period.

They used the previously introduced animal model (by Henry, 1992) who showed in an experiment with mice that "Methyl Prednisolone" could significantly reduce Noise Induced Hearing Loss (NIHL) only at the highest tested frequency of 48 kHz and had no significant effect at other tested frequencies of 3, 6, 10 and 24 kHz. Methyl prednisolone was injected (40 mgkg) 15 min before noise exposure from a digital noise generator and four additional doses were given at 8 hours intervals.

The only treatment that has proven to be effective in the treatment of sudden sensorineural hearing loss is the steroid drug Prednisone. A short course Prednisone will, in majority of patients, improve the recovery of their hearing. The amount of recovery that can be anticipated is based on a variety of factors, the most important of which is how severe the hearing loss is. It is important that the prednisone be administered as soon as possible after the onset of the hearing loss as we have only a limited amount of time during which the hearing loss remains reversible. (Cited in http:// www. ent,utmem. edu/vestibulococh/shl. html).

Prednisone is a very effective immune suppressant, however, it does have side effects. Prednisone, may cause alteration in mood, with some patients feeling either depressed or very euphoric It may cause water retention and stomach upset. When used for a longer period of time, it is always combined with a medication to protect the stomach from forming ulcers. Prednisone may cause significant increases in blood sugar in patients with Diabetes and must be used with extreme caution in patients with this disorder.

Long term prednisone therapy may cause bone wasting. A very rare complication of prednisone therapy is a condition known as a vascular-necrosis of the hip which can cause serious damage to the hip joint. (Cited in http://www.ent.utmem.edu/vestibulococh/shl.html).

Exploration of the middle ear with repair of an inner ear fistula is recommended in patients with a clear history of sudden hearing loss associated with diving, straining, attitude change, or recent otologic surgery. The role of surgery in patients who do not improve with non-surgical therapy remains controversial. (Cited in http://www.ent.utmem.edu/vestibulococh/shl.html).

In summary, sudden sensorineural hearing loss remains a poorly understood and frustrating problem for the otolaryngologist. More rational treatment will probably only follow the elucidation of the specific cause or causes of this disease process. (Cited in http:// www.ummah.net/software/softbase/sudden.txt).

MANAGEMENT OF SUDDEN DEAFNESS

PAST MANAGEMENT APPROACHES

The management of sudden cochlear hearing loss, unrelated to recognizable specific ear lesions, has been based entirely upon idiopathic and presumptive etiologic concepts and on empiric therapeutic theories. Presumptive diagnosis of vascular cochlear accident, viral disease, endocrine, autoimmune or allergic causes have been made and empiric nonspecific treatment has been advised. Single or multiple modalities have been in use, including vasodilators, steroids, hyperbaric oxygen therapy, vitamins, intravenous procaine, intravenous histamine, adenosine triphosphate, other drugs and surgical procedures, such as stellate ganglion blocks used on empiric grounds. No single definitive therapeutic approach has been in general use. On the contrary, so called "shot-gun therapy" has been and continues to be normative. In recent years most investigators have been stressing the greater value of steroid therapy in most idiopathic varieties of sudden hearing (Goodhill, V., & Harris , J (1979))..

Spontaneous Recovery and the Treatment Evaluation Dilemma

Among the difficulties in assessing both presumptive diagnosis and empiric therapies are the frequent spontaneous recoveries noted in many patients who have had no treatment. Such recovery may be partial or total and may occur within a matter of hours, days, weeks or months. Controlled statistics comparing untreated and treated

patients are virtually nonexistent. In idiopathic syndrome cases, the etiology must usually be considered presumptive, and only the idiopathic label is realistic. Objective etiology can be confirmed surgically in some labyrinthine membrane rupture syndrome cases, but even in such cases as in idiopathic syndrome cases, the final definitive answer must await postmortem temporal bone-histopathological studies. (Goodhill and Harris (1973)).

Only in surgically explored labyrinthine membrane rupture syndrome cases does some objective information begin to emerge regarding etiology and therapy. When an otosurgeon sees a definitive oval or round window fistula or both, in sudden hearing loss cases, and monitors the results of surgery by otologic audiologic studies, it becomes possible to introduce some objectivity into etiology and therapy. Such data are by no means sufficient for evaluating the total labyrinthine membrane status, which must await adequate postmortem temporal bone histopathologic studies. Even marginal objectivity is lacking in the evaluation of etiology and therapy in the larger idiopathic syndrome group. (Goodhill and Harris (1973)).

The Present Management Approach

The current management approach is based upon an evaluation of the history and findings. Every patient with a sudden spontaneous cochlear hearing loss is considered to be an otologic emergency. (Goodhill and Harris (1973)).

Management of Mild to Moderate Sudden Sensorineural Hearing Loss Cases

All patients with mild to moderate losses (30-50 dB SRT, 40-80% SDS) are placed at bedrest at home with head elevated 30 degree and allowed out of bed only for necessary attendance at the clinic for otologic and/or general medial diagnostic studies. No physical stress (exercise, sexual activity, walking or sports) is allowed. Only bathroom privileges are allowed. No initial medication other than necessary tranquilizers and sedatives are advised during the work up period. In every case of sudden sensorineural hearing loss, complete otologic, radiologic, vestibular and internal medical studies are necessary. Every attempt is made to find a specific otologic cause such as chronic osteamostoiditis with fistula, suppurative labyrinthitis,internal auditory meatus or cerebellopontine angle lesions, previous ear surgery, ear trauma or head trauma. If a definitive otologic cause can be elicited appropriate management can be carried out. If no such otologic cause can be elicited, thorough general medical studies are carried out to elicit possible factors such as viral, vascular drug and other "idiopathic" medical causes. Occasionally, unsuspected problems such **as** clotting defects, bradycardia, auricular fibrillation, vascular occlusions and other definitive general medical conditions are discovered. If there is no suggestive labyrinthine membrane rupture history, the idiopathic syndrome is suspected, and the presumption is made that there is a relationship between the recognized general medical condition and the hearing loss and the appropriate medical therapies are instituted. In those patients in whom ENT and/or general examination suggests recent viral or bacterial

upper respiratory infection, empiric broad spectrum antimicrobial therapy is started and changed, when possible, to specific antibiotic therapy, if pathogenic bacterial organisms can be cultured from the nasopharynx, nose or sinuses. If there is no medical contra-indication a course of empiric Adreno-cortico Trophic Harmone or Prednisone therapy may be started. (Goodhill and Harris (1973)).

The patient is asked to avoid physical exercise in spite of an apparent systemic etiology. Such systemic lesions may or may not be etiologically related to the sudden sensorineural hearing loss lesion. Conversely a specific history of barotrauma or physical stress does not necessarily prove.a labyrinthine membrane rupture etiologic relationship. When very definitive circulatory upper respiratory diseases, or other possible general medical conditions are discovered and appropriate treatment is instituted, prompt beginning improvement in hearing may be noted in a few days. General supportive therapy includes high vitamin B and C intake, avoidance of tobacco, caffeine, alcohol, salicylates and the common allergenic foods (milk, chocolate, nuts, shellfish) that might contribute to membrane edema. Audiometric evaluations should be done every 3-4 days to monitor hearing levels. If significant serial audiometric gain is demonstrated, this conservative therapy is continued until audiometric findings show a return of hearing to within normal limits. Thus, if the hearing return reaches a level of 20-25 dB with 70-80% SDS, no further treatment is considered necessary, unless the intensity of vertigo and/or tinnitus continues. Such clinical improvement following nonspecific management may not be accompanied by a precise final etiologic diagnosis. (Goodhill and Harris (1973)).

If there is no spontaneous improvement in hearing in a mild to moderate case of idiopathic syndrome following 7-8 days, a course of empiric corticosteroid therapy is advisable. If the hearing does not remain stable but continuous to drop in both SRT and SDS measurements, and/or if there is increased tinnitus intensity and/or if increased vestibular symptoms occur, the management should be changed as follows:(Goodhill and Harris (1973)).

Management of Unresponsive or Severe Total Sudden Sensorineural Hearing **Loss Cases**

Idiopathic Syndrome cases

Those patients with idiopathic syndrome with severe or total losses (80-100 dB SRT, 0-30% SDS) or those with moderate hearing loss who are nonresponsive and show no improvement on empiric medical therapy, including corticosteroid therapy and bed rest at home for 7-10 days are hospitalized and placed at absolute bed rest with no lavatory privilages with head elevated at 30 . The course of corticosteroid treatment is continued till completion. If there is no beginning hearing gain as measured by bedside audiometry after 3-4 days of such hospitalization (total of 12-14 days), surgical exploration should be considered, in spite of the absence of history suggestive of labyrinthine membrane rupture.

In cases of labyrinthine membrane rupture with severe or total loss (80-100 dB SRT; 0-30% SDS), the patient is immediately hospitalized and kept at absolute bedrest with head elevated at 30 degree. Hospital medical consultation is secured in the search for possible unrecognized vascular, viral or other systemic diseases. Daily

bedside audiograms are done. If the labyrinthine membrane rupture diagnosis appears tenable on the basis of a significant physical stress history, and if the general medical history and the findings are completely negative, the likelihood of membrane rupture is considered to be significant and early surgical exploration is indicated. No absolute guidelines can be laid down for the timing of the surgical exploration, but it usually should be considered within the first 10-12 days following the physical stress episode.

Rehabilitation

Young patients who do not recover spontaneously from a unilateral sudden deafness should have preferential seating in the classroom and be advised on maneuvering persons in conversation to the side of their better ear. Children and adults must be acquainted with their inability to localize the source of sound and must truly stop, look and listen rather than just listen, when crossing the street. The use of amplification with the contralateral routing of signals (CROS) hearing aid may benefit some of these patients (Harford and Barry, 1965; Harford and Dodds, 1966).

Those patients who do not recover serviceable hearing from bilateral sudden deafness should have speech reading and auditory training to enable them to make the most of their residual hearing. Appropriate amplification with a hearing aid should be utilized when appropriate. (snow, J.b. (1973)).

GLOSSARY

Adenoca rcinoma: Carcinoma derived from glandular tissue or in which the tumor cells are from recogniz -able glandular structures. Adeno- carcinoma's may be classified according to the predominant pattern of cell arrangement, as papillary, alveolar etc. or according to a particular product of the cells as in mucinous adenocarcinoma.

Atrophy: Wasting away, a diminution in the size of a cell, tissue, organ or part.

Arteritis: Inflammation of artery.

Cardiopulmonary: Pertaining to the heart and lungs.

Chlamydia A genus of bacteria of family chlamydiaceae, order chlamydiales, occurring as gram negative, coccoid orga- nism that multiply only within a host cell and have a unique growth cycle. They are common pathogens of animals and cause a variety of diseases in humans. Called also as PLT group and formerly Bedsonia, Chlamydozoon and Miyagawanella.

Cogan's Syndrome Non-syphylitic keratitis with vestibulo auditory symptoms.

Concussion A violent jar or shock or the condition which results from such an injury.

Cryptococcus A genus of a sexual yeast like organisms of the family CRYPTOCOCCAE - which usually have a capsule and do not form a pseudomycelium as do the candidae.

Cyclosporine A cyclic peptide produced as a metabolite by the soil fungus Tolypocladium-inflatum-gams that selectively inhibits activation of helper T lymphocytes; used as an immuno -suppressant to prevent rejection in organ transplant recipients.

Cytomegalovirus Any virus of the subfamily Betaherpes virinae, highly host specific herpes virus that infect man, monkeys or rodents with the production of unique large cells bearing intranuclear inclusions. Depending on the age and immune status of the host, cytomegalovirus can cause a variety of clinical syndromes collectively known as Cytomegalic Inclusion Disease, although the majority of infections are very mild or

subclinical. Called also as ' SALIVARY GLAND VIRUS' and 'HUMAN HERPES VIRUS-5". Abbreviated CMV.

Dimer i) A compound formed by the combination of two identical simpler molecules.

ii) A capsomer having two structural units.

Diuretic An agent that promotes the excretion of urine; increasing the secretion of urine.

Erythema A name applied to redness of skin produced by congestion of capillaries, it may result from a variety of causes.

Fibrin The insoluble protein formed from fibrinogen by the proteolytic action of thrombin during normal clotting of blood. Fibrin forms the essential portion of blood clot.

Fibrinolysis The dissolution of fibrin by enzymatic action.

Gastric Pertaining to, affecting or origination in stomach.

Granulomas Imprecise term applied to any small nodular delimited aggregation of mononuclear inflammatory cells.

Herpes A spreading cutaneous eruption, any inflammatory skin disease caused by Herpes virus and characterized by the formation of clusters of small vesicles. When used alone, the term may refer to Herpes simplex or to Herpes zoster.

H.Simplex A group of acute infections caused by herpes simplex virus type 1 or type 2, characterized by the development of one or more small fluid filled vesicles with raised erythematous base on the skin ormucous membrane, and occurring as a primary infection or recurring because of reactivation of a latent infection. Type 1 infections usually involve nongenital regions of the body whereas in type 2 infections, the lesions are primarily seen on the genital and surrounding areas, although there is overlap between the two types. Precipitating factors include fever, exposure to cold temperature or to UV rays, sunburn, cutaneous or mucosal-abrasion, emotional stress and nerve injury.

Hypercogulable Characterized by abnormally increased coagulability (susceptibility of becoming clotted).

Hyperostosis Hypertrophy of bone; exostosis.

Hypertrophy Enlargement or over growth of an organ or part due to an increase in size of its constituent cells.

Hypokalemia Abnormally low potassium concentration in the blood, may result from excessive potassium loss or by the renal or the gastrointestinal route from decreased intake or from transcellular shifts. It may be manifested clinically by neuromuscular disorders ranging from weakness to paralysis by electrocardiographic abnormalities, by renal disease and by gastrointestinal disorders.

Hypoplasia Incomplete development or under - development of an organ or tissue; it is less severe in degree than aplasia.

Immune Mediated Immunity (protection against infectious disease conferred either by the immune response generated by immunization or

previous infection or other non-immunologic factors) mediated by lymphocytes either through release of lymphokines or through exertion of direct cytotoxicity transmissible by transfer of lymphocytes but not serum; it comprises delayed hypersensitivity reactions; systemic response to viral and microbial infections, contact dermatitis, granulomatous reactions etc.

Interferon Any of a family of glycoproteins that exert virus non-specific but host specific antiviral activity by inducing the transcription of cellular genes coding for antiviral proteins but selectively inhibit the synthesis of viral RNA and proteins. Interferons also have immunoregulatory functions (inhibition of B cell activation and antibody production, enhancement of T cell activity & enhance -ment of NKcell cytotonic activity) and can inhibit the growth of non-viral intracellular parasites. Production of interferon can be stimulated by viral infection, especially by the presence of double stranded, RNA, by intracellular parasites, by protozoa and by bacteria and bacterial products (endotoxins).

Interferon can be divided into 3 distinct types (£, β and) associated with specific producer cells and functions, but all animal cells are able to produce interferons and certain producer cells (leukocytes and fibroblasts) produce more than one type (Interferon £ and Interferon β) Abbreviated IFN..

Lassa Fever

An acute febrile disease caused by Arena virus (Lassa fever virus), endemic through out west Africa and spread by contact with multianimate rats (Mastomys nalatensis) which sheds the virus in its urine, or by interpersonal contact, most infections are subclinical or mild, although severe cases resulting in death occur, symptoms include fever of insidious onset, headache, dry cough, backpain, vomiting, diarrhoBa pharyngitis, facial edema and occasionally a maculo popular rash; in severe cases there is a sudden drop in B.P. on the seventh day, with death resulting from shock, hypotension, peripheral vasoconstriction, hypovolemia and anuria. Sensorineural deafness which may be permanent, sometimes results.

Leukemia A progressive malignant disease of the blood forming organs, characterized by distorted proliferation and development of leukocytes and their precursions in the blood and bone marrow.

Meningitis-Meningococcal An acute infectious disease attended by seropurulent inflammation of the membranes of the brain and spinal cord and due to infection by Neisseria — meningitidis. The disease usually appears in epidemics and symptoms as those of acute cerebral and spinal meningitis usually accompanied by an eruption of cutaneous eythematous, herpetic or harmorrhagic spots. The malignant form is known as **"Water House -Friederichsen syndrome".** Called also cerebrospinal fever and Epidemic cerebrospinal meningitis.

Metatarsus The part of the foot between the tarsus and the toes, its skeleton being the five long bones (metatarsals) intending from the tarsus to the phalanges.

Monoclonal Drived from a single cell; pertaining to **a** single clone.

Mononucleosis Presence of an abnormally large number of mononuclear leukocytes (monocytes) in the blood. The term is often used alone to refer to infectious mononucleosis.

Mumps An acute infections disease caused by a paramyxovirus, spread by direct contact, airborne droplet nuclei by infectious saliva and perhaps urine and usually seen in children under the age of 15; although adults may be affected. Usually associated with painful swelling of one or both parotid glands; other salivary glands may also be involved.

Mycoplasma A bacterium of Class "Mollicutes".

Myeloma A tumour composed by cells of type usually found in bone marrow.

Plasmogen Activator A general term for a group of substances that have the ability to cleave palsminogen (in active form of plasmin) and convert it to plasmin its active form.

Pleotropy The quality of a gene to manifest itself in more than one way i.e. to produce more than one phenotypic expression.

Rubella An acute, usually benign infectious disease caused by a Togavirus and most often affecting children and nonimmune young adults in which the virus enters the respiratory tract via droplet nuclei and spreads to the lymphatic system. It is characterized by a slight cold, sore throat and fever followed by enlargement of the post auricular, suboccipital and cervical lymph nodes and the appearance of a fine pink rash that begins on the head and spreads to become generalized. Transplacental infection of the fetus as a result of maternal infection in the first trimester can cause death of the conceptus or severe developmental abnormalities in the newborn infant. Called also as German measles and three day measles, and Rubeola in French and Spanish.

Scarlet fever Infection with group A, β-hemolytic streptococci of varying severity, although it usually has a milder cause than in the past, where septic complications such as otitis media, mastoiditis and suppurative lymphadenitis are not rare. It is characterized by pharyngitis . and tonsillitis concurrent with a typical erythemamous

rash, produced by an erythrogenic toxin elaborated by the streptococci, progressive from the trunk and neck to the extremities (except the palm and soles) forehead and flushed face and circumoral pallor, red/ white strawberry tongue and lines of hyperpigmentation (Yastia's sign) in the body creases; the rash disappears and is followed by desquamation of the skin. Similar clinical manifestation, but usually with involvement of pharynx and tonsils may follow injection of wounds, burns of the skin with group Aβ-heamolytic streptococci that elaborates an erythrogenic toxin. Called also as "Scarlatina".

Schwa nnoma : Neoplasm, originating from schwann cells (of myelin sheath) of neurons.

Seroconversion The change of a serologic test from -ve to +ve indicating the development of antibodies in response to an infection or immunisation.

Serum Clean portion of any body fluid, the clean fluid that separates from blood on clotting is called blood serum.

Spuma virus Foamy viruses; a genus of non-pathogenic viruses of subfamily spumavirinae (family Retroviridinae).

Steroid A group name for lipids, includes progesterone, bile acids, sterols (such as cholesterol), toad poisons etc.

Systemic Pertaining to or affecting the body as a whole.

Temporal Pertaining to the lateral region of the head, superior to the zygomatic arch; pertaining to time.

Titer The quantity of a substance required to produce a reaction with a given volume of another substance, or the amount of one substance required to correspond with a given amount of another substance.

Tuberculosis Any of the infectious diseases of man and animal caused by Mycobacterium, characterized by the formation of tubercles and necrosis in the tissues.

Vertebrobasilar Pertaining to or involving vertebral and basilar arteries.

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