SUDDEN DEAFNESS : A REVIEW

REG. NO: 8705

AN INDEPENDENT PROJECT WORK SUBMITTED IN PART FULFILMENT FOR FIRST YEAR MASTER OF SCIENCE (SPEECH & HEARING) TO THE UN.VERSITY OF MYSORE

ALL INDIA INSTITUTE OF SPEECH & HEARING

MYSORE-57O0 06

MAY 1988

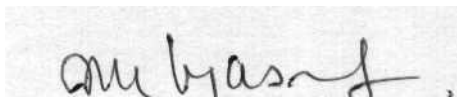
To

My beloved Parents, Sisters

and Brother,

C_E_R_T_I_F_I_C_A_T_E

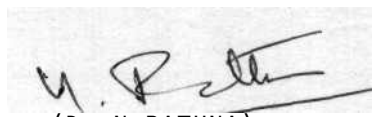
This is to certify- that this Independent Project entitled "SUDDEN DEAFNESS A REVIEW" has been prepared under my supervision and guidance.



(Dr.M. N. Vyasamurthy
Guide.

C_E R T_I_F_I_C_A_T_E

This is to certify that the Independent Project entitled **"SUDDEN DEAFNESS A REVIEW"** is the bonafide work on part fulfillment for the degree of Master of Science (Speech and Hearing) of the student with Register No.3705.



(Dr.N.RATHNA)
DIRECTOR
All India Institute of Speech & Hearing,
Mysore.

D E C L A R A T I O N

I hereby declare that this Independent Project entitled "SUDDEN DEAFNESS" is the result of my own study. Under the guidance of Dr.M.N.Vyasamurthy, Department of 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. Reg.No. M. 8705

Dated: May 1988.

ACKNOWLEDGEMENT

I express my sincere gratitude to Dr.M.N.Vyasamurthy for his invaluable guidance and help in undertaking this project.

I extend my thanks to Dr.R.Rathna, Director, All India Institute of Speech and Hearing, Mysore.

I would like to express my thanks to Mr.Vasuki, Miss.Rohini, Miss.Manjula, Mr.Vasu, Miss.Asha for their encouragement, Co-operation, understanding and timely help in completely my project.

My special thanks to Mr.Prasad for his encouragement and moral support.

Last, but not the least, my sincere thanks to my brother Mr.Venkatesh for taking such great care in typing out my project.

CONTENTS

C H A P T E R - I

INTRODUCTION

The sudden onset of severe or total hearing loss is a most dramatic event in the life of the person who is so afflicted. Prosper meniares Was the first to report about a case with sudden idiopathic sensorineural hearing loss refers to the occurance of an abrupt loss of hearing with no discernible cause. Pars inferior which ia a portion of the inner Ear, including cochlea and saccule, is more susceptible to various injurious agents. These factors are responsible for acute injury that results in sudden deafness. Sudden hearing loss may also be present with any disorder of the external, middle ear and in some occasions with the disease of the otic capsule of neural connection.

The severity of the hearing loss in sudden deafness may vary from mIld to total loss of perception of the most intense sound. The loss may be permanent or temporary. One of the constant problems in any study concerning sudden hearing loss iS the fact that on occasions

there is a sudden return of hearing or a spontaneous improvement without any treatment at all.

Some of the definationa given by different authors haa been quoted below.

Sudden hearing loss can be defined as a mild to total loss of hearing of sensorineural type, which is most often unilateral and develops over a period of few hours or few days. (Lindsay '59)

Goodhill ('7l) defined sudden deafness as one whose development is sudden or instantaneous or rapid which is usually unilateral loss and is unrelated to middle ear disease in patients with presumably unknown previous ear or hearing problems.

Saunders ('72) uses the term sudden deafness to describe "a unilateral, generally abrupt loss of hearing of idiopathic type". Siegel's ('73) defined sudden deafness as "an abrupt hearing loss of sensorinanral Type".

Sudden sensoryneural loss is a frightening experience for the patient. The patients worse fears concerns tumours or stroke. The physician is aften frustated by a symptom whose cause at times is not apparent. Every effort has to be made to identify the causes of sudden hearing loss.

Sudden deafness is a disease of multifactrorial genesis. The cause remains as even unknown. Proposed explanations for sudden hearing loss as listed by Simmons include meningitis, encephalitis, sickle cell crisis, mumps, parotitis, multiple sclerosis, syphilis, cogans disease, acoustic neuroma, meniere's disease, head injury, sludging of blood, thrombosis, spasms, toxic drugs, ear surgery, rupture of oval and round window, viral and bacterial infections.

The symptoms of sudden deafness are varied and often fluctuate. In many individuals, the hearing loss is first noticed on awakening in the morning. Most frequently associated with sudden hearing loss are such auditory complaints as tinnitus which is noted in about

70 - 85% of individuals and is often described as changing in pitch. In about 50% of persons, the hearing loss is preceded, accompanied or followed by vertigo, dizziness or feeling of unsteadiness. Some patients may report a feeling of fullness or plugging in the affected ear. On rare occasions when sudden idiopathic sensory neural hearing loss is bilateral, both ears may be affected simultaneously or the second ear may become affected months or years later.

The pathogenesis of sudden deafness has been partly understood for most etiologies except a few conditions like viral disorders, which have been batter understood. For conditions like vascular disorders which cause sudden deafness the explanation is that a thrombosis or an embolism may obstruct some of the important vessels supplying the end organ, leading to a total or partial obstruction of blood flow to the region which in turn may lead to sudden hearing loss.

Tha pathology of the endorgan has been studied in detail by Schuknecht ('62) and others. They reported that the organ of corti was missing in some parts, the hair cell and the ganglion cell population was highly decreased. The tectorial membrane, reissne3s membrane and stria vascularis ware all collapsed. There wag degneration of the saccule also.

Audiological finding in sudden deafness present a variety of pictures, in different cases. The only general aggrement in the audiogram is that all cases appeared to be sensorineural. The degree of loss may vary from mild to moderately profound. Maximum speech intelligibility scores vary depending on the site of auditory disorder.

Impedance results characteristically show normal'A'type tympanogram with normal static compliance results. In individuals with profound hearing loss reflex thresholds cannot be measured

because signals cannot be presented at sufficiently high intensity levels. If the site of lesion is cochlear the findings include absence of tone decay, presence of recruitment on ABLE, high SISI scores and type 'I' Bekesy tests.

Sudden deafness has generally been regarded as a medical emergency and patients have most commonly been hospitalized immediately. Sudden hearing loss has for its treatment as many varied therapies as there are in most of medicine's unsolved problems. It is obrious that since the precise cause cannot-be determined in moast cases a specific method is like wise absent.

Some of the treatment methods for sudden hearing loss include harmone therapy, steroid therapy, vasodilators, de-congestants, nicotinic acid, heparin, cortisone, vitamin therapy, carbogen inhalation, suggestion therapy, dehydration therapy and

surgical treatment. Most of the treatments are synptomatic.

Although a large number of treatment methods exists for sudden deafness, it is very difficult to say which is the best, as the spontaneous recovery of hearing to normal or near normal level in these patients make revolution of any form of therapy very difficult.

- x - x -

C H A P T E R - II

CHAPTER-II

HISTORY AND ASSOCIATED SYMPTOMS

One of the most outstanding feature of sudden deafness is its suddan onset of symptoms. The most common presenting symptoms which is reported most frequently is tinnitus (Snow'73, Morrison '75, Grase '76). The tinnitus vary in degree i.e., it may develop hours or days after the deafness has appeared, or come on simultaneously with the problem (Snow '73) or it may either proceed the hearing loss (Morrison '75). The quality of tinnitus is usually tickling type, but Jaffe has also reported tickling bell like noises. The duration of persistence of the tinnitus may vary from one month to many years, ever last the deafness. However Mironeuko ('76) Schuknecht ('76), Goodhill ('72) reports tinnitus as being a very frequent symptom in sudden deafness. There are dual views regarding the presence or absence of tinnitus as being an indicative of prognosis.

Morrison ('75) says that prognosis is not affected by the presence or absence of tinnitus on the other hand Fredreick('79) is of view that the presence of tinnitus is an indicative of poor prognosis.

Vertigo is an incomistent symptom seen in sudden deafness (Sownders '7l) About the incidence of vertigo, mild or transient vertigo is seen in about 40% of the patients incapacitating vertigo which lasts for 4 to 7 day is seen in 10% of the patients. Thus the incidence of vertigo vary considerably Joseph('75) reports that the vertigo may accompany or follow hearing loss by hours or days or even years. It can also be the only symptoms in some cases.(Goodhill '79)

There is a considerable amount of support given by a study (Morrison '75) about the relation between the frequency of vertigo and the site of the pathology. Acute vertigo is often a prominent

symptom in patients with an end organ localization or sub total deafness. It is less often to be seen in truly sensorineural leison and in retrocochlear cases it is more unusual. (Semory 71%, Total or sub total - 66%, semorlneural - 37%, Neural - 10%)

Cochlear leisons whan they do not respond well to treatment then vortigo do affect the prognosis. As reported by Squnder's ('77) the symptom of vertigo with a down ward sloping audiogram was a significant prognostic factor if the deafness accompany the vestibular disturbances. However vertigo forms an irrelevant factor for the prognosis in patients with a flat or upward sloping audiogram.

In any case, Yashida ('7l), Nomer ('72), Hozawa etal ('76) and Emagi ('76) have reported of occurrence of vertigo with sudden deafness.

DEAFNESS AND AUDIOLOGIC STUDIES:

As already mentioned tha symptoms of sudden deafness is

sudden in nature and the evolution of those symptoms present a great deal of variations. In case of sudden onset, a semation of loud sound is usually seen in the affected side. This is the first sign most often reported by the patient with sudden deafness hearing loss is seen only in the next stage, which may develop over the course of an hour, a day or several days associated with cracking or popping sensation and this annoying tinnitus sometimes mistaken for a stroke by the patient.

Usually this sudden deafness is noticed first by the patients when he gets up in the morning after he had gone to sleep in the night with perfectly normal hearing. In some cases they may get up in the midst of their sleep due to the annoyance caused by the tinnitus. Simmons ('77) in his study on 89 patients

with sudden Hearing loss reported the presenting history of the patients ag follows - present on awakening - 33, exertional - 24, sendentary - 15, eating - 5, not instant loss - 5.

It was also noted that hearing loss were rarely seen during physical or emotional strain or at rest which are the usual day to day life activities.

Usually a person with sudden deafness experience difficulty in telling exactly when the hearing loss was noticed. This is more true in cases with unilateral hearing loss since it escapes their notice untill a situation arises where there is a need for using both ears to the maximum extent. However loss can be noted promptly in bilateral hearing loss cases since it comes in the way of inter-personal communication to a large extent.

These people are able to detect their hearing loss inspite of presence or absence of tinnitus. Since they experience profound loss of discrimination initially. They also experience difficulty in localizing sound since the unusal sensitivity to intense sound and diplacusis are rare in these cases.

Audiometric findings in these cases reveal flat type of audiogram in pure tone audionetry, + ve SISI, + ve ABLB, - ve tone decay, poor speech discrimination as seen in SN loss, Type II or Type III in Bekesy audiomotry. However involvement of vestibular function are rare in these cases.

In case of pure tone audiometry even though audiogram shows different pattern general uniformity is observed. Where sudden sensory losses show flat type of audiogram or show a high tone

cut-off; high frequency loss seen in sensorineural loss.

Morrison '75 reports of retrocochlear has having low-tone, trough-shaped, flat or high toned audiogram. Recruitment is usually absent in the initial stages but once loss becomes permanent then it can be noticed. (Snow '73) Van Dishoeck '57 and Verge '61 reports of the presence of recruitment in less than 50% of the patient with permanent loss. The audiometric patterns recorded by Shaia etal ('76) based on his study in 1220 cases is as follows; Flat - 32%, High tone - 51%, Profound or total loss - 25%, Low tone - 12% .

According to a study by Jerger ('60) type III Bekesy tracings was reported in the majority of patients with permanent hearing loss of sudden onset. Shaia etal ('76) reported the findings in 1220 cases of sudden hearing loss. The Bekesy patterns

as recorded by them are as follows; Type I - 22%, Type II - 59%, Type III - 11%, Type IV - 8%.

In a study by Jerger ('6l) two third of the patients with permanent hearing losses of sudden onset had low SISI scores, Type III Bekesy audiograms, ran measurable discrimination scores and minimal recruitment which are usually associated with a neural hearing loss and one third showed high SISI scores, Type II Bekesy audiograms, some recruitment and maeasurable but low discrimination scores which are associated with cochlear hearing loss. Flarbert '64 based on the above results divided the loss into cochlear and neural.

Graham etal in his study on 30 patients made use of the factors given by Saunders ('77) on the audiometric findings in localizing the site of leison as cochlaar or retrocochlear. Electro-

cochleographic results agreed with the diagnosis in 23 patients and thus confirmed their diagnosis in all 25 patients.

Neill ('60) reported of a typical pattern of audiometric test response, obtained with cases of sudden deafness associated with mumps. BC thresholds were characteristically lower than AC thresholds speech reception thresholds were higher than the average pure tons loss, discrimination scores were very low and there were indications of recruitment.

Based on the andiologic test results, Beeningham ('62) has differentiated between the sudden loss occuring due to presence of high intensity noise and the sudden onset of hearing loss due to explosion, or sound shock. The earlier one is always unilateral and is characterized by a flat curved, puretone

audiogram with + ve recruitment. The second one is a high frequency loss with no vestibular vertigo accompanying it.

Depending upon tha vestibular symptom Kirikae etal ('64) divided their group of sudden deafness patients into too, the Ist had high tone loss, with directional changing, positional nystagmus and the second group had hearing loss over the whole range with directional hystagmus towards one side. This difference was supported to be due to quantitative factors of a lesion in the vestibule.

Stephens etal ('67) reported a patient in whom a type IV Bekesy audiogram was found during the acute phase, with the other tests indicative of neural lesion, but the type IV Bekesy audiogram then converted in Type II within 5 months with the

other tost results indicating a cochlear lesion. The authors concluded that the different patterns of response to auditory stimuli were associated with different stages of recovary following sudden deafness while the hearing loss is stabilizing. Altshuler & Welsh ('69) aho reported the same such variability of these special audiologic tests with time in a given patient has been observed by others.

Vestibular signs are evident in the majority of patient with vertigo and in a few with no vortiginous symptom. Approximately 50% of the patients vith sudden deafness will have some permanent abnormality of response of the vestibular apparatus to caloric stimulation. Abnormalities vary from complete loss of response to mild variations from normal (Snow '73).

Pain is an occasional feature in sudden idiopathic deafness like, the pain some times experienced by patients with acoustic neuroma, it is a dull mastoid or ear ache. It is more likely to

be seen in patients in whom there is a neural element to the hearing loss Morrison '75). A section of pressure in the affected ear is experienced by many of these patients (Snow '73). Headache is occasionally encountered and symptoms of viral upper respiratory tract infections occur as frequently as in 25 percent of the patients in some series. Fever, unusually 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. Byl found elevated while cell count in 59% and elevated erythrocyte sedimentation rate in 41% of the cases with sudden deafness.

Age and sex distributions are fairly equal. Sudden semory deafness is possibly common in males. The ages of these patients range from adolescences to 72 years (Morrison '75) with 30 to 60 years being the most crocial range (Simmons '73) Nadol ('75) reports that sudden deafness occurs most often between 8th to 20th years of age Shaia ('76) in his study of 1230 cases of sudden deafness concluded that there were slightly more females than males, although the difference was very small. Three - fourth's of his patients were 40 years of age or older at the onset of the symptoms.

Morrison ('75) also reported that the retrocochlear lesions were common in the younger age group. He also felt that in discussing post viral sudden deafness, children are difficult to categorize since they were usually investigated months or years after the onset.

Seasonal incidence has also been carefully examined in sudden deafness. No pattern has emerged to support a viral etiology. Between '63 to '75 with a number of scattered exceptions, sudden deafness had started in every calender month of every year and no more than 5 cases had commenced in the same month of the same year (Morrison '75).

Singleton etal ('77) evaluated 51 patients suspected of having a perilymph fistula. They postulated that predominantly vestibular complaints had unrecognised perilymph fistula. Positional nystagnus was found to occur in supine, right down and head hanging right positions. This Nystagmus had little or no latency and lasted for long and did not change its direction on sitting up. Vestibular lesions with abnormal caloric responses are also a common finding in

patients with mumps deafness (Welsh '63) and (Hyder '79) and in acoustic neuroma.

Kinkawada ('79) has reported of a long term follow-up study of unilateral sudden deafness patients. It was performed in 28 cases who ware diagnosed as having sudden deafness 10 years earlier. 18 cases showed no change on audiogram, 5 cases showed deterioration of hearing in the affected ear. In 2 cases the affected ears had remained unchanged while the unaffected ears showed deterioration of hearing. Bilateral progressive hearing loss was found in 5 cases. None of the patients with vertigo at the onset of sudden deafness had vertiginous attacks later. Those patients who had showed marked recovery usually maintained good hearing. The author has thus stressed that audiometric follow up are highly essential in these cases for differential

diagnosis.

SPONTANEOUS RECOVERY:

In cases with Psycbonic deafness the occurrence of spontaneous recovery is probable and it occurs frequently in cases with multiple sclerosis. In cases of definite viral infections such as Zoster, mumps, and measles, even though the recovery may not be complete spontaneous recovery do occur to some degree. In case of ototoxic drugs such as salicylate and quinine reversibility is their feature. Minimal improvements in the auditory thresholds are likely to be seen in cases with vascular leisons, without any specific treatment. When it comes to sudden loss due to hydrops only a small percentage of patients recover spontaneously, whether hydrops are due to syphilis or other diseases of the otic capsule or it may be idiopathic.

Secretory otitis media being a very common cause of sudden deafness in all age groups, there is a possibility of missing the diagnosis without typanogram. Spontaneous recovery occurs in such occasional sudden sensory deafness. This diagnostic difficulty can be avoided if the stapedial reflex is used for localization purposes (Morrison '75).

Morton etal ('66) have reported the case of an old lady whose deafness recovered spontaneously without the use of drugs. The testings revealed that the loss was predomenantly of the neural type. The authors have hypothesized that the probably etiology is a vascular insult.

Singleton ('7l) stated that 25% recover completely, 25% show some improvement and 50% have a lasting profound hearing loss. Schecides ('7s) considers that in 40 -60% of patients

with sudden deafness we can expect a spontaneous recovery.

Mashs etal ('78) reported that spontaneous recovery could be expected in 65% of the patients. Those patients with an upward slope of audiogram have a recovery rate of 92%, downward slope 28%, and if loss is worst near the base of the cochlea, i.e., 8 KHz it is 25%.

PROGNOSIS:

In cases of sudden deafness approximately in one third of patients hearing returns to normal limit and one third are left with a 40 to 80 dB SRT, and ona third have total loss of useful hearing (Snow '75). Thus the prognosis for the recovery of the hearing in sudden deafness is not so bad as is generally supposed. Shimozaki ('74) in his study followed up cases of

sudden deafness where the following factors ware analyzed - symptoms, sox, age, method of treatment, audiogram types and prognosis. Complete recovery was seen in 11%, clear recovery in 16.7%, slight recovery in 33.5% and 53.9% remained unchanged. In conclusion he made an emphasis on an "Early treatment" since the possibility of hearing improvement was found to be limited to one month from the onset of hearing loss.

Symptom of severe vertigo in patients with a downward sloping audiogram is a significant prognostic factor. Failure of hearing recovery is strongly associated with severe Vertio (Fortbild '65). Rubin {'68) found that prognosis depended upon severity, none of his severe cases showing any improvement.

Snow ('73) postulates that prognosis cannot be easily related to the rapidity of onset, the presence of tinnitus

- recruitmant or type of Bekesy audiogram. The longer the the delay between the onset of deafness and the onset of recovery the worse the prognosis for complate recovery. But Simmons ('73) is of view that prognosis correlates best with the interval between symptom onset and the first audiogram and very poorly with either the type of treatment or the interval between symptoms and the patients first visit to a physician. Spontaneous recovery to normal hearing is more likely to occur if the deafness is not associated with severe vertigo and if deafness is not total initially (Snow '75).

Morrison ('75) is of the view that prognosis is poor in sudden total deafness, not very favourable in sensory and sansorineural hearing loss, but excellent in neural leisons.

Hiraki etal ('76) performed electrocochleography on 34 patients with sudden deafness. The various wave form patterna which they got were (l) action potential high response (2) decreased action potential high response (3) action potential low response (4.) dominant - ve summating potential (5) + ve or - ve summating potential (6) action potential and summating potential no response.

Fifteen patients had complete recovery. The ecochgram showed dominant - ve summating potential wave forms in 8 of them, and action potential high response wave form in 7.

In 9 cases there was slight improvement. Here the findings were; Action potential low response in 2; +ve summating potential in 4, - ve summating potential inl; action potential and summating potential no response in 2.

In ten patients of no change. Action potential and summating potential no response was seen in 7 cases. - ve summating potential in one and decreased action potential high response in two cases.

So the authors concluded that ecochggrams can be used to estimate the prognosis of sudden deafness in initial stages. Similar results haw also been reported by Nishida ('77).

Most authors also agree that early cases do well and that the chance of recovery decreases with increasing time after onset, this hypothesis is typified.

Hypo active caloric responses predict a very poor prognosis for higher frequency recovery and a normal caloric response may correspond with a better prognosis (Simmon '77). Nevertheless complete recovery of hearing does occur at times

even after several weeks of profound loss. Yoshida etal ('77) have used equilibrium test to detect prognosis. The absence of gaze nystagmus and normal caloric response had good prognosis.

Some of the features not favourable for prognosis are increasing age, diabetes, hypertension. Severe vertigo, late treatment, downward sloping audiogram, elevated sedimentation rate, delayed diagnosis. (Frederick '77, Simoon's '77 and Shaia etal '76)

Feature which aids in better prognosis, are time between onset and diagnosis, hearing at 8 KHz, better speech discrimination scores, absence of vertigo and early treatment (Simmon '77, Shaia etal '76).

Anthony's '78) finding that, in a large treated series,

the recovery rate was 87% for those seen at three days or less, 75% at up to one week, 53% at up to one month, 16% at up to three months, and 10% if seen and treated at over three months from onset similar findings have been reported by Shaia etal ('76).

The audiogram curve is also found to carry some prognostic valve (Emmett etal '79) feel that an upward slop or flat audiogram carries a better prognosis than a downward sloping audiogram. Poor prognosis is also related to those having an initial audiogram which reveals a conplete loss of 8 KHz. Speech discrimination valve is found to have no prognostic significance. Simmons ('77) says that the threshold at 8 KHz carries a prognostic significance. If loss is either improving or stable at 8 KHz, the prognosis ia good or complete recovery is seen in 78% of cases. If there is no hearing at 8 KHz, regardless of hearing at other frequencies,

prognosis is 29%. A flat hearing loss has a better prognosis if the threshold is no worse at 8 KHz than is at other frequencies. A worse prognosis is there is an additional loss at 8 KHz.

C H A P T E R - III

CHAPTER-III

C_A_U_S_E_S

Several causes can account for the sudden onset of hearing loss. In about 30% of cases a clear cut cause can be determined. The cause of the sudden hearing loss in a patient is identifiable from the history. However estimates of these cases classified as obscure, idiopathic or unknown range about 10% to 22% for example. Occlusion of the vasculature of the inner ear with subsequent hearing loss in one of every thousand pUmp-by-pass surgeries. Acoustic neuroma. accounts for 1% to 2% of al1 sudden hearing loss.

Hallberg's report in 1956 discussed 178 cases of sudden hearing loss. He listed 9 classes into which he had been able to distribute the patients.

1. Vascular accidents.

2. Unknown.

5. Honiara's disease.

A. Toxic effects on the labyrinth from other infections.

5. Emotional episodes, including malingering.

6. Acute rise in labyrinthine pressure.

7. Injection of an orthobiotic serum.

8. Neuritis of eighth cranial nerve.

9. Systematic diseases.

Stahle.J and Oberg in 1960 believed that the sudden deafness was due to cochlear oedema.

Schuknecht in 1962 provides evidence which suggests that mumps or mumps like virus may be an important factor

in causing sudden deafness. He examined the temporal bones of 4 patients with unilateral sudden hearing loss. The changes were similar to those which occur in human labyrinthitis of known viral etiology and are unlike the pathological changes resulting from experimental vascular occlusion in animals. So he suggested that viral infection should be considered as important etiological factors for sudden deafness.

Newby in 1964 thought that sudden deafness was due to interruption of blood supply to the cochlea, probably a spasm of the cochlear artery. If the condition persists for more than a few hours cochlea suffers irreperable damage and the hearing loss becomes permanent.

Carco in 1967 says that circulated disturbances can

cause sudden deafness either cochlear or transcochlear in nature.

Jaffe in 1967 gave reasons for S.H.L. as complications of viral infections, vascular disorders, metabolic toxicities, trauma post-operative status, pregnancy and also a large group where no cause is recognised.

Simmons in 1968, Morrison in 1971 and Schiff in 1974 gave encepalitis as a cause for sudden deafness. Encephalitis has probably a viral origin which affects the substance of the brain. Sudden deafness is one of the results along with other problems caused by encephalitis. ECG tests shows that the patient will have normal puretone audiogram, yet no understandings of speech. The same thing develops over a period of weeks and months featuring headaches,drowsiness, epilepsy and abnormal

movements. In tho beginning steges it is often confused with psychogenic deafness.

Joseph in 1975 gave syphilis as a reason for sudden deafness, be it congenital or acquired.

Incidenca of sudden deafness as reported by him are as follows.

Late congenital syphitis - 18%

Early latent syphilis - 17%

Late latent syphilis - 25%

Symptomatic neuroma - 80%

Asymptomatic neuroma - 25%

The clinical picture in both early and late forms of congenital syphilis are similar having sensory neural loss with low discrimination scores occuring bilaterally.

The reason that ototoxicity causes sudden hearing loss was given by Morison in 1975; by drugs like neramycin, streptomycin. Kanamycin and neomycin are said to alter the cochlear functions. Kanamycin damages haircells which is followed by neural damage. The damage of haircells which starts in the basal coil progresses apically.

Also; Morrison in 1975 said that 3% of the patients with menier's disease were prone to get deaf unilaterally. The second ear may get affected after an interval of months or years.

In 1978 Richars, experimentally showed that a total vascular occlusion and anoxia too leads to sudden hearing loss. A concussion type head injury with anoxia and or haemorrage can also be the etiologies. The patients level of consciousness can be used to

guide us about the intensity of sensorineural deafness.

David in 1980 described acoustic trauma as a cause for sudden deafness. Boenninghams ('62), Kevata and Suga ('87) and David say that a noise induced deafness occurs suddenly after a period of exposure to uniform loud noise.

Maures in '62 and Robert ('69) have also reported acoustic trauma to be a cause for sudden hearing loss. Some rare cases have also been observed of which electric shock, shoulder pains, inhalation of nitric oxide and typhoid fever are sone, reported by Gaillard ('66), Nakagawake ('77), Mack)'76) and Morrison ('75) respectively.

After having listed out the known causes of sudden deafness we will now come to the "unknown" causes of sudden deafness.

Three theories have been name (l) Viral infection

(2) Vascular occlusion (4) Membrane rupture.

A list of proposed causes of sudden sensorineural hearing loss has been given below.

1. Viral infection:

a) Viral cochleitis

b) Viral labyrinthitis

c) Viral neuritis, auditory nerve

d) Viral polyneuropathy - including Ramsay-Hunt -syndrome

e) Viral induced meningoencephalitis

2. Cochlear membrane breaks:

a) Intracochlear breaks

1) Reissnar's membrane tear, with and without hydrops (theore

2) Rupture of spiral ligament

b) Oval window and round window membrane tears

1) Secondary to head injury

2) Secondary to congenital malformations

5) Post stapedectomy

4) Compression - decompression

5. Vascular occlusion:

a) Complete thrombus or embolus of labyrinthine or cochlear artery - microcmboli secondary to routine or pump-bypass surgery.

b) Partial:

1) High viscosity syndromes: macroglobulinemia, polycythemia vera

2) Small-vessel obstruction: sickle-cell anemia, microemboli,bubble (Caisson's disease)

5) Small-vessel narrowing diabetes mellitus, atherosclerosis, Buerger' s disease

4.) Hypercoagulability states

5) Vasospasm

c) Inner-ear hemorrhage: leukemia, anticoagulated states

A- Bacterial infections:

a) Meningitis, encephalitis, labyrinthitis secondary to chronic ear infection or surgery

b) Syphilis, primary through tertiary

5. Autoimmune_disorders:

a) Inner-ear autoimmune disease

b) Relapsing polychondritis

c) Polyarteritis nodosa

d) Cogan's syndrome

c) Sarcoidosis

6. Neurologic disorders - multiple sclerosis:

7. Neoplasms:

a) Vestibular schwannoma

b) Metastatic cancer

8. Ototoxic drugs (bilateral loss)

VIRAL INFECTION:

Severe haircell losses; strial atrophy and atrophy of the organ. Of corti are noticed when histopathological studies of patients are carried out. Known viral infection and such as herpes zoster oticus measles, mumps etc., are clearly perceived by the mind by through histological findings. Though studies have shown an increase in the number of cases due to viral infections those particularly said to cause sudden hearing loss are mumps, varicella zorter, rubella, herpes simplex. Wilson found that herpes virus family was unique in the sense that herpes infections that were associated with an S.H.W. occured as a part of multiple

viral infection in 70% of cases which is a characteristic not acquired by other naurotropic virus.

The list of viruses associated with sudden deafness as put forward by Jaffa ('78) are.

VIRUS		CLINICAL SYNDROME
Mono virus	-	Upper respiratory infection
MycoplaSma	-	Mumps
PasainfluEnza (type 3)	-	Measles
Herpes hormines	-	Herpes Zoster.

Many theories have come up but it is still not clear whether there exists a definite cause and effect relation between upper respiratory infection and sudden deafness. SD is said to be caused by mumps on the onset of parotitis.

Mumps may cause (a) unilateral total deafness

(b) partion deafness mostly in children.

(c) Severe bilateral deafness.

Though uncommon, Lindsay and Menenway ('54.) and Morrison ('75) have reported of sudden deafness in measles.

Herpes Zoster is associated generally with facial paralysis and very rarely with sudden hearing loss.though the patient expeniances painful vesicular eruptions in the outer ear. Polio viras, german measles, yellow fever, cytomegalo virus infection have also been associated with sudden deafness.

VASCULAR OBSTRUCTION:

Partial or complete occlusion of the inner ear vessels have been said to be the cause of sudden hearing loss. Experiments conducted on guinea pigs caused an irreversible loss of the cochlear microphonic within one minute. In acoustic neuromma surgery to a similar condition occurs. Unintention trauma, to the labyrinthine artery also results in irreversible immediate

sudden deafness.

Patients undergoing cardia-bypass surgery are also prone to acquire sudden deafness.

But in general the fact that occlusion causes sudden hearing loss is unsatisfactory.for example temporal tone histopathologic findings for those patients with sudden hearing loss do not resemble those seen in patients with innar ear vascular obstruction or haemorrage.

The other different conditions leading to sudden deafness of vascular origin are listed as vasospasms, thrombasis, hyper-coagulation and sludging of blood. All the above conditions in general result in a rapid complete and irreversible hearing loss.

COCHLEAR MEMBRANE BREAKS:

Hypothesis say that membrane tears poison the auditory sensorineural structures with nourotoxic, pottassium rich endolymph thus causing fluctuating hearing loss.

Cochlea membrane breaks may also occur at the oval or round windows or both of them. The fluctuating hearing loss and dizziness is owed to perilymph breakage from the inner ear through the break or fistula.

Compressive, decompressive episodes, head injury or stapedectomy may also cause membrane breaks resulting in sudden onset of deafness.

Recent reports cite that the perilymphatic vessels, the

stria vascularis and the integrity of the cochlear dect might be involved in the genesis of sudden hearing loss.

- xx - xx -

C H A P T E R - IV

C_H_A-P_T-E_R- IV

H I S T O L O G I C A L S T U D I E S

Histological studies of cochlea which have suffered sudden hearing loss are relatively rare. Lindsay (1959) reported one case of degenerative changes in the stria vascularis. Schuknecht, etal (1962) found some loss of haircells but variability in the presence of damage to supporting structures in different specimens. They also discovered changes in the organ of corti which ranged from what was described as 'shrinkage' to total loss of the organ. In these and other cases, they report changes in the tectorial membrane and in the stria vascularis.

Beal ('67),Joseph ('75), Tomoyukl ('78), Lindsay ('73), Nomura ('76), Isamo and Sando ('77), David ('57) reported that ganglian cell population is decreased at the nasal turn but being normal towards the apex. Reissensrs membrane would be collapsed and adherent to the basillar membrane. Saccule would be involved,

while utricle and the semi circular canals escaped any damage.

Schuknecht's ('73) report of pathological changes of eight temporal bones in sudden deafness cases is given in the table.

ORGAN OF CORTI

Specimen	Hair cells	Supporting cells	Tectonal membrane	Stria Vascularis	Other changes
1. A.P.		+ + +	+ + +	+	atrophy of sacculo
2. B.H.	+	+			Otoscleroais
3. M.T.	+++	+ + +			-
4. B.W.L.	+ + +	+		+ + +	Otosclerosis
5. B.W.R.	+ + +	+		-	Large cochlear aqueduct. Saccular enlargement.
6. H.H.		+	+ + +	+	-
7. P.C.	+ + +	+ +		+ + +	Large cochlear aqueduct.
8. G.S.	+	+ + +	+ + +	+ + +	-

By performing electrocochleographic studios on patients with sudden deafness it was thought that the pathophysiology of this disease could be deduced. The various patterns of AP & SP responses obtained in cases of sudden deafness were classified into the following types.

a) AP high response.
b) Decreased AP high response.
c) AP low response.
d) Dominant - SP.
e) - SP or + SP
f) AP & SP no response.

The cases showing the type of dominant - SP & AP high response had satisfactory prognosis. In these cases the sensory epithelium and the cochlear nerve seemed to indicate a reversible conditions being affected by the temporary functional block. Further more, it seemed to indicate that the neural regions related to the source of AP (N_1) response were impaired in cases

in which CM were recorded at normal response. In the unsatisfactory prognosis eases with decreased AP high response, AP low response and AP, SP no response in which only the extremely low or depressed CM responses could be recorded it seemed that the sensory epithelium and the cochlear nerve were affected permanently although the degree of impairment varied (Nishada.H. '78)

Kiniura and pearlman ('56 & '58) have reported of experimentally evaluated histopathological changes which are slightly different from the one reported by clinical studies. Here the pathologic changes involved more severe damages of the ganglion than hair cells, with little variation in different turns of the cochlea. There was loss of spiral ligament cells, variable loss of hair cells. There was little effect on tectorial membrane. Permanent and complete arterial occlusion produced generalized destruction of the inner ear structure s.and fibrous tissue invasion and finally complete ossification of the cochlea.

Alford ('65) and Suga ('70) also come with similar reports.

Hoshino ('78) examined, under a scanning electron microscope, the left ear of a 57 year old female, Mho suffered sudden deafness during the course of relapsing polychondritis. Found marked degeneration of the organ of corti in all turns and dislocated and encapsulated tectorial membrane in the cochlea. Marked decrease in number of sensory cells in the utricular and sacular maculae and total loss of sensory hair bundles in the ampullary cristae of the semicircular canals were seen in the vestibule. These findings strongly suggests that the cause of sudden deafness in this case might be viral.

Grundfast.K.M.('78) said that inner ear fluid dynamics and patency of the cochlear aqueduct appear to be important factors in pathogenesis. According to Blair Simmons ('78) change in fluid pressure in one intracochlear compartment also causes a pressure

gradient in the other. Thus one membrane break is likely to cause a secondary break. He presents indirect evidence for such double membrane breaks in sudden deafness and argues that one of the two breaks can heat spontaneously while the other remains active.

While the knowledge of pathological changas in sudden deafness is still limited, temporal bone studies in mieniers disease provides important information on its pathogenssis. Many authors have described enlargment of the cochlear duct as evidence for endolymphatic hydrops. Maximum enlargement of the endolymphatic system disrupts the membrane system which seperates endolymph from perilymph and can be confirmed by histologic examination. This indicates that a change of the ionic concentration in the inner ear fluids is responsible for inner ear dysfunction (Ilberg '76).

Ottsaki.K. ('80) conducted comparitive study of serum protein in patients with sudden deafness. He found that the mean of serum total protein in the sudden deafness group was significantly

lower than that of normals. (P less than 0.05)

Several significant histopathological findings were noted in a case of sudden hearing loss in a patient with chronic lymphocytic leukemia. The major pathological findings were leukemic haemorrage into both perilymphatic and endolymphatic spaces in the cochlear and vostibular systems endolymphatic hydrops in the cochlea and sacculus and a relatively narrowed and straightened vestibular aquiduct and endolymphatic sac. Additional interesting findings include loss of hair cells in the organ of corti and vestibular end-organ, destruction of the striavascularis possibly the origin of the blood, fibrosis in the perilyrmphatic spaces in the cochlea and the vestibule and in the endolymphatic space in the vestibule. The leukemic infilberate observed in both the cochlea and the vestibule was not considered to be significant. (Isamu '77)

Sando.I ('77) presents the histopathological study of

two cases of sudden deafness. The temporal bones showed cochleasaccular abnormality. The most striking pathological changes were collapse of the organ of corti, atrophy of the tectorial membrane, atrophy of the stria vascularis reduction in the number of cochlear nerves, collapse of the saccular membrane and partial absence of the sensory epethelial layer in t he saccular macula. These changes are quite similar in type to those occuring in labyrinthitis of known viral etiology. In addition, unusual findings of endolymphatic hydrops limited to the extreme basal end of the cochlear duart ware found in case.l. A patient cochlear aqueducb and circumscribed perllymphatic labyrinthine ossification in the superior semicircular canal were also observed with those histopathologic findings, the possibility of viral infection via the meninges as well as via hemotogenous route into the inner ear is proposed.

PATHOGENESIS:

There are three routes of access of viral particles to the inner ear. The most common is viremia, in which the virus are deposited within the membranous cochlea. The second route is where viras gain access to the inner ear through the perilymphatic Space from the subarachnoid space via the cochlear aqueduct. Finally it is the direct extension from middle ear to inner ear. (Lindsay '59 and Snow '75)

Once the virus particles enter the membranous cochlea, pathophysiological start occuring which are often reversible, but once the destruction is extreme, the loss is permanent. Vascular endothelium is invaded first and inclusion bodies may be sean in the endothelial cells. Hyperamia occurs at the site of inflammation. When virus particles attach to erythrocytes, sludging of blood and hemaglutination occurs. Oedema in the perivascular spaces occur when there is infilteration of different cells. This may lead to

necrosis of neuroepithelium. (Snow '73)

Transmission of viral infection from brain to labyrinth has also been reported by Erich ('76). Here there is an infilteration by lymphocytis and histocytes along the nerves in the internal meatus into the spiral ganglia and the scalatynpani (Lindsay '59). There is an infilteration of various cells like histocytes, lymphocytes, plasma cells into the inner ear which leads to fibrosis and then some sensory cells disappear.

It has been unable to isolate viruses of any type in viral antibody studies, C.S.F. studies, postnatal swab studies or by the paul-bunnell test. Morrison ('75), Jaffe ('67), Shaffer ('64), Welsh & Welsh ('65). Alteration of the immune response in the grand state, antigen-antibody reaction, or hypersensitivity have all been reported as the cause of sudden deafness, in conditions where pregnancy leads to sudden deafness. There is a high occurrence of false positive, biological test results like VIRL, CWR or RPCET,

which may be temporary or chronic. (Morrison '73, Jaffe '67)

Kenneth ('75) report that ottco barotrauma is a frequent cause of perilymphatic fistula. It may also result from rapid alterations in perilymphatic fluid pressure. But for this to occur the cochlear duct should be patient or there should be an underlying middle or inner ear congenital abnormality. (Parisel and Bukea) Kannath '73) says that the eustachian tube is lined by a surface active membrane which has a surface Tension lowering properties similar to pulmonary surfactant. It allows the lumenal wall of the tube to seperate in response to the pull of the musculature. It gets destroyed by proteolytic enzyme producing bacteria pressure equalizing capacity of the eustachian tube is affected. In these conditions ottico baro-trauma occars. When the air pressure build up in the eustachian tube is abnormal cochlea may be injured and there will be inward and outward displacement of the stapes foot plate in the oral window, creating pressure distortion of the membronous labyrinth. In these conditions besides

pressure, haemorrage from torn vessels and gas bubbles released can also damage the cochlea. (Berkley '70)

Goodhill and Simmons '71) expiation for the occurance of sudden deafness in cases of perilymphatic fistula is due to rupture of labyrinthine window membrane due to pressure exerted on it. This rupture in turn leads to exertion of pressure on reissners basilar and tectorial membrane, which may rupture if pressure is great, which leads to mixture of perilymph and endolymph. So partly hearing loss is due to actual tissue injury at the site of break and partly due to mixture of fluids.

C H A P T E R - V

C_H_A P_T_E_R- V

T R E A T M E N T

Sudden hearing loss has for its treatment as many varied therapies as there are in most of medicines unsolved problem. It is obious that since the precise cause cannot be determined in most cases, a specific method is likewise absent. Treatments appear and disappear and then are rediscovered at irregular intervals.

One of the constant problems in any study concerning sudden hearing loss is the fact that on cccassions there is a sudden return to normal hearing or a spontaneous improvement without any treatment at all. This thereby increases tha difficulty of attempting to prove the therapeutic efficacy of any Medicament. .

Sudden deafness has generally been regarded as a medical emergency (Jaffe) and patients have most commonly been hospitalized immediately. Reports indicate that recovery is relavitaly rapid if it were to occur. Sheely (1960) reports that 62% of his cases recovered when treatment was given within 4 days and 47% recovered when treatment began within six weeks. So it seems to be clear that severer the hearing loss and the larger period from the disease onset to the start of the treatment, the poorer the

recovery of hearing (Guilton'65)

Cases of sudden deafness have a very little tendency to relapse, at any rate, not, within a short time. However a few cases of relapse have been reported, one of them after an interval of 15 years. To improve the balance of neuro-vegetative system by dietary measures and anti-allergic or endocrine treatment, the physician can try to improve the deficient properties of these constitutionally predisposed patients. (Guilton '65)

The therapies currently advocated includes vasodilation, steroids, anticoagulation, reduction of the viscocity of the blood, sedation and tranquilization, vitamins, corticosteroids, bedrest etc. One of the main problem for selection of therapeutic method is etiology. It is important to know whether the cause of the problem is classified under 'known' or 'unknown' group.

Under the cause 'unknown' group treatment methods diverse because the data are confusing and the etiology is in doubt. There will be a controversy about the natural history. Treatment

is empirical and effectiveness of some treatment methods has been doubted. (Richards '78)

In the group where the cause has bean established (Syphilitic tuberculosis and acute inflammatory disease of the nervous system, neoplasms including acoustic neuromma, meniere'a disease etc.) the treatment problems are those of the individual theraphy for the condition. In proven viral infections, Morrison etal (70) find it right to advocate the use of steroids but it haa not been found to be of much use in cases of deafness due to measles or mumps which is of longstanding nature.

Vasodilation has been advocated by Van Dishoeck and Bierman ('57), Sheely ('60) Jaffe ('67) Rubin ('68) and Newby ('64)

The results of vasodilation therapy appear to approximate spontaneous recovery rate (snow '75). The rationale of vasodilation therapy in view of the preponderance of evidence for the viral etiology for sudden deafness is questionable. Now vasodilation is advocated in view of the vascular

changes that occur in viral diseases.

Study of the effect on cochlear blood flow in guinea pigs by vasodilating agents was done by Suga & Snow, who showed that nicotinic acid, even in massive doses, has no messurable effect on cochlear blood flow. Histamin Phosphate and Betahistamine increased cochlaar blood flow in dosages that produce bronchospasm in guinea pigs and may well produce vasodilation on the basis of anoxia.

In case of sudden severe hearing losses the prompt administration of a vasodilator, perhaps in combination with an anticoagulent will sometimes result in a dramatic restoration of hearing. From time to time, there has been reports in the literature concerning the beneficial effects of vitamin therapy in improving the hearing, but the consensus of medical opinion is that sensory neural hearing loss is irreversible. (Newby '64.)

Osterwald & Bouche ('73), Barton ('74.) these authors also concludes that vasodilator therapy supplemented by cortesona, antibiotics and vitamins is highly succesful especially in cases treated early.

Jaffa ('67) advocates low molecular weight dextran to reduce the viscosity of the blood. It is administered by the intravenous route in 500 cc quantities over a period of 4 hours. Dextran is a plasma volume expander and increases cardiac output and vascular profusion. It also reduces blood viscocity, platelet adhesiveness, sludging and cochlear formations with resultant improvement of micro-circulation. It is an asset to patients with sudden deafness.

Rossberg.G ('77) evaluated 2 methods for treating the affli-cation of sudden hearing loss. The effects of treatment with dextran 40 and papaverin ware compared with those of a polypragmatic drug administration. The latter proved to be effective in cases of sudden hearing loss, however more significant improvements were obtained when papaverin and dextran 40 were administered. Polypragmatic therapy did not influence the hearing loss in cases of morbus meniere, in contrast, with papaverin/dextran 40, considerable increase in hearing could be demonstrated.

Kellerhals.B ('77) and Schatzle ('81) reported that dextran infusion can be used as the treatment of choice for cases of sudden severe hearing loss and acoustic trauma.

Morrison and Booth ('70) have drawn attention to the importance of early steroid treatment for sudden deafness. Authors have basically used Prednisone. A dose of 30 mg daily for the first week was found to be adequate. Higher doses were used without obvious benefit. Improvements in the puretone audiogram within normal limits during the first week was found in most and only rarely after 10 days. During the second week the dose of predisona was reduced to 25 mg and thereafter to 20 mg daily. Steroids wore withdrawn if there was no betterment after 2 weeks. If there was a dramatic improvement, withdrawal was spread more slowly over several weeks to avoid the possibility of relapse. Pregnancy diabites and hypertension have been contraindicated.

Wilson. W.R. ('80) conducted double-blind studies for the treatment of idiopathic sudden hearing loss with oral steroids. He found that steroids had a statistically significant effect on the recovery of hearing in patients with moderate hearing loss.

Villar.J. ('8l) did statistical review of 60 cases of sudden idiopathic neurosensory deafness of probable vascular or viral etiology treated with nicergoline, corticosteroids and in 18 cases, diatrizoate meglumine. Total or partial auditory recoveries were obtained in 36% of the cases; in the cases with association of

diatrizoate maglumine, the percentage of recovery was 38%. Steroid harmone (Predonine) administered to patients with low-tone deafness of sudden onset resulted in the recovery of hearing to normal levels shortly after treatment. (Yagi.T.'82)

Four patients with sudden deafness were treated with steroids immediately after the initial symptoms of sudden deafness. Hearing loss of all cases became worse in a period of 2 or S days after the steroid administration. Their hearing acuity was markedly impaired. However, the steroid therapy was continued and the hearing loss showed the remarkable improvement after 7 to 20 days of treatment. Minami.Y (82') concludes that if the patient has major side effects there is no necessity of cessation of steriod administration even when the hearing loss was progressive after the steroid therapy was started.

Some of the newer cortico steroids which have been used in the treatment of sudden deafness are dexamethasone, parametha-sone, triamcidone, methylprednisolone or tetracosactrin. If the lesion is shown to be sensory or sensorineural, vasodilator therapy has on occasion been instituted in addition to the steroid therapy. Nadol ('75) has reported chance of improvement in 50% of the cases with syphilitic deafness.

Steroids with anticoagulants have also been used like the adreno cortico tropic hormone (ACTH). Bolognesi ('60) used ACTH with a case of hyper coagulation and found complete recovery. Saunders has noted the uae of ACTH in the viral and vascular etiology of sudden hearing loss, where in viruses attach themselves to red Mood corpuscles and produce henagglutination and has reported improvement.

ACTH has maty functions. It stimulate the production of cyclic AMP from ATP. AMP causes a decrease in aggregation of platelets. It also exerts a beneficial effect of beta (vasodilator) versus alpha (vasoconstrictor) catecholamines. ACTH is a stimulant for corticosteroids from the adrenals. It tends to suppress antigen-antibody reaction. The union of antigen-antibody is not prevented, nor is the release of hostamine from sensitized cells prevented. ACTH has a distinct lipolytic effect, actuated by a specific lipase enzyme system. This helps clear the blood of chylomia by permitting the triglycericles to be reduced to glycerol and free fatly acids which are then metabolized.

The mechanism of action of ACTH on its receptor cell is to stimulate the formation of adenyl cyclase which is a messenger

from tha endocrine gland to the cell interior. This in turn releases the amount of cyclic AMP from the ATP that is found within the cells. By increasing the amount of cyclic AMP and ATP, there is a general increase of ADP available. This has an effect on the aggregation of platelets. Since platelet aggregation is increased by the presence of ADP, there is an increased tendency for sludging and rouloua formation with occlusive effect on the small vessels in the periphery. ADP has a characteristic of increasing the stickiness of platelets. Ishigama etal ('76) Jakobi etal ('75), Meyerhooff ('79) have all given evidence to support the recommendation of ATP in the treatment of sudden deafness.

Inhalation of pure O_2 mixed with 5% CO_2 gas has been used as tha therapy in cases of sudden sensorincural hearing loss. Three patients, in whom hearing was lost 44, 25 and 45 days respectively, after the occurance of sudden deafness and without improvenent in hearing, showed a significant improvement in hearing after inhalation of this gas mixture. He also reported that this therapy was effective for patients observed soon after onset of hearing. (Muruta.K ('78)

Flsch,V ('85) reports that in patients with sudden deafness , the o2 supply to the vestibular tissues is significantly reduced but the response to carbogan in sudden deafness is not accompanied by a reduction (stealing effect), but by an increase of perilymphatic Oxygenation. Therefore carbogen inhalation was used for the treatment of sudden deafness. In a prospective randomized study carbogen inhalation yielded significantly better results than the intravenous infusion of papaverine and low-molecular dextran. Carbogen inhalatio is recommended for the effective, non-invasive treatment of sudden deafness.

The techniqua of hemodilusion was assessed in 12 cases of sudden hearing loss as it is known to improve peripheral circulation. Good results were obtained in 9 patients. The immediacy of the effects suggests the real efficacy of hemodilusion as opposed to spontaneous recovery. Hemodilusion is supposed to improve oxygenation in the cochlea. This technique is very simple and quick to perform. (Dauman.R.etal ('83)

The other drug to gain importance in the treatment of sudden deafness is Heparin. Bolognesi ('60) suggested Anticoagulation as a possible method of treatment in the very early cases of sudden deafness of obscure etiology. He placed each patient on heparin and warfarin. He observed complete recovery of hearing in 3 out of 5 patients.

Schiff.M ('74) discusses the use of heporin as excellent agents in producing prompt recovery in sudden deafness cases. These drugs tend to inhibit or ameliorate vasculitis, inhibit hypercoagulati and decrease hyperlipidemia. These effects encompass the rationale of its use in sudden deafness.

Piccoli.A ('77) reports results with a new heparinoid (3 GS) in the treatment of recent and longstanding sudden loss of hearing. This study suggests that good results will be obtained if 3 GS ig given immediately after onset. The drug is not contraindicated and is completely free of side effects. Donaldson ('79) used

heparin with 25 cases of sudden deafness and found that 14 had complete recovery (60.4%), 2 had good recovery (8.7%), 7 had poor recoveiy (50.4%). None of the patients failed to recover.

The inhibitory effect of heparin on antigen-antibody reaction was noted by Drugstedt. This mechanism is related to the fact that haparin complexes and binds histamina quantitatively. This in turn prevents the capability that histamine had of carrying the sodium ion across a cell membrane such as a fibroblast, thereby damaging the internal Mechanism of the call. This binding of histamine and heparin limits the cytodestructive effect to the benefit of the capillary vith it3 endothelial cells with other responsive cells.

Heparin specifically stimulates Up protein lipase formation. This engine is located in or near the vascular wall. The lipolytic effect is observed at a considerably lower concentration than that used to prolong clotting time. Spencer's showed that a decrease of the lipoprotein factor in blood effected an improvement in hearing.

Kuglar.R ('73) considers that the stellate ganglion block is a method of choice in the treatment of sudden deafness. Jocobson ('74) has also used stellate ganglion block (N=10) and drops of complamin in chaomacrodex in his therapy for sudden deafness. Stellate ganglion block is made up of inferior cervical ganglion and the superior thoracic ganglion of the sympathetic narve trunk. The function of this trunk along with the rest of autonomicnarvous system is to assist in preparing the body to meet emergency i.e. for fight. This is accomplished by constriction of blood vessels, increased blood pressure, enlarged pupils etc. Plester in sixties was the first to use this in treatment of sudden deafness.

Some of the complication of stellate glanglion block as put forward by Haug etal ('76) are penumothorax, temporary loss of voice due to trauma of the recurrent laryngeal nerve and temporary dysphagia from trauma of vagus nerve.

Simmons.F.B. ('73) reported that the treatment with Mannitol for one week gave spectacular results and the hearing on the effected side returned to normal. Pang.L.Q.('74) presents 2 cases of sudden sensorineural hearing loss following diving, and the succesful treatment by recompression with complete recovery of the hearing.

Microwave was used in the treatment of patients with sudden deafness by Kawamoto.H. ('76). A patient was irradiated with 20-50 watt microwave apparatus at frequency of 2450 MHZ from exterior auricle on the affected side towards the inner ear once or twice a day. Microwave irradiation was considered to improve blood flow in such arteries as vertebral basillar anterior, inferior cerebellar labyrinthne and thus do better the condition of the affected inner ear.

Emmett Etal (1981) discusses the action of hypaque in sudden hearing loss. He examined 16 patients complaining of sudden hearing loss and tinnitus who met the criteria of no vertigo, and a hearing loss that was not complete ware administered vasodilator therapy

including hypaque. 15 patients had complete return of their hearing to pro sudden hearing loss levels. 2 patients had partial return of their hearing, but not up to serviceable level and ona patient had no response to treatment. Shea.J.J. ('78) also used hypaque with vasodilators with sudden hearing loss patients of less than one month duration and no vertigo at onset. He found 86% of the patients had return of serviceable hearing.

Four cases of sudden deafness, healed by the use of trans-4 aminomethyL-cyclohexane carboxylic acid (tranexamic acid) was reported by Ohsaki.K.etal ('76). Tranexamic acid is an antiplasmic agent. Therefore, it was presumed that tranaxamic acid could act effectively on such phenomena as acceleration of permeability as tissue edema and exrtravascular blood oozing as the result of hypoxia followed by sludging in inner ear.

Fielder.H.('84) treated 33 patients with sudden hearing

loss with Natrium-Meglumine-diatrizoat (Urografin), When all other standard therapies failed. Detailed investigations were carried out and thorough measures were taken against anaphylactic reactions before and during the intravenous administration of Urografin. In 10 cases (30%) an improvement of hearing of atleast 20 dB was observed with restitution of almost normal hearing in some cases. The treatment was most succesful in those cases where an excessive hearing loss (more than 40 dB) occured for the first time. Since the drug is only intended for diagnostic use further Use as a therapeutic agent will require legal endossment.

Recent laboratory and clinical studies provide increasing evidence that idiopathic sudden hearing loss can be effectively treated. First, corticosteroid therapy has been shown to be efficacious in restoring hearing in patients with moderate hearing losses. Secondly, studies of cochlear blood flow and perilymphatic oxygen tension indicate that positive modifications in $P0_2$ can be

obtained with 5% CO- and 95% oxygen. Future clinical trials are likely to show that improved oxygenation of the cochlea will have a positive effect upon hearing recovery. Third, patients with a clear history of perilymph fistulas are managed by middle ear exploration and repair (Wilson '84).

Vestibulatory is one another surgical procedure which has boon recommended to patients of sudden deafness due to labyrinthine window rupture by Fiedeman ('75).

The less popular mathods for treatment of sudden deafness are use of methylgluoanine salts of which belong to the triohenzoic acid derivatives. (Fukuoka ('78), injection of amino-triovate-intra-venously (Mortimitsue '74-). The last of the treatment methods is the uae of ultrasonic's. (Kazama '76)

- X - X -

B_I_B_I_L-I-O G R_A_p_H_Y

1. Altshuler.W.Morton & Welsh.Z.0liver; "Sudden hearing loss with spontaneous recovery". Journal of Speech & Hearing Disorder Vol.31, 166-171, 1966.

2. Ammar - Khooja.A -"Sudden deafnass following a scratch induced benign dymphoreticulosis". Rev.Laryngol. Otol. Rhinology 105 (l) 1982, 63.

3. Anderson.R.G., Meyerhoff.H.L. "Sudden sensorineural hearing loss." Otolaryngol.Chin.north. Am 1983 Feb.l6(l) 169 - 95.

4. Anon - "Treatment of sudden deafness with radire puerarial with comparitive analysis of therapeutic results in 294. cases". Chinese Med.Journal. 10, 1973, 591 to 595.

S* Armstrong, B.W.; "Sudden Deafness in unoperated ear after gtapedactomy"; Archives of Oto laryngology. Vol.86; 156-159; 1967.

6. Arnold. W. & Vosteen, K.H.; "Sudden deafness as a consequence of rupture of basilar artery;" HNO Vol.25(4); 127-130; 1977.

7. Balkany. T.J. "Reversible sudden deafness and vertigo" EENT Mon, 55(5), 1976, 148-151.

8. Barton R.P.E.; "Sensory neural deafness of sudden"onset"; British Journal of audiology. Vol.8: 119-120; 1974.

9. Beal,D. David, Hemenway, G.Williams and Lindsay.R.- "Inner ear pathology for sudden deafness"; Archives of Oto laryngology; Vol.85; 591-598; 1967.

10. Bergstrom,La. Vonne, Baka, B.Bruce and Sando Issmuko; "Sudden deafness & Facial palsy from metastatic bronchogenic earcinoma"; Journal of Laryngology & Otology-.Vol.91(9); 787-793; 1977.

11. Bhandari.R. Steirman.G.S. "Sudden deafness in Chiokenpox, a case report". Ann.Neurol. 1985. Mar.13 (5) 347.

12. Bolognesi A.V.B.. "Sudden deafness - Five cases treated with anticoagulants"; Archives of Otolaryngologlea. Vol.72; 31-40; 1960.

13. Bolognesi.A.V.B. - "Sudden deafness."Archives of Otolaryngoly 72, 47-55, July 1960.

14. Bosatra - A.B. & G.B.De Stefani. "The idiopathic sudden - deafness. Clinical study". Acta Otolaryngol. Supp, 169(1961)

15. Bouche. J. etal. "Sudden deafness" Prof.Actuals (RL 1973,7-42.

16. Carco.P. "Sudden deafness of vascular origin". Int. Audiol, 6, 1967, 72-75.

17. Chapman.P; "Naproxen and sudden hearing loss". Journal of Laryngol. Otol. 96(2) 1982, 162 to 166.

18. Currier.W.D.: 'Metabolic errors and sudden deafness". Otolaryngol.Chin.N.Amer. 8(2), 1975, 501-505.

19. Dauma.n.R. Aran J.M. - "Parallel evolution of audiometric and Electrophyaiologic thresholds in sudden deafness". Rev.Laryngol. Otol. Rhinol - 1982, 103 (5) 377-80.

20. Dauman.R etal; "Hemodilution in. sudden deafness first results" Arch. Oborhinolaryngol. 1983 238(2) 97-102.

21. Emmett. J.R. "Diatrizoale meglumine (Hypaque) in treatment for sudden hearing loss" J.Laryngol. otol Suppl. 4, 1981 139 to 142.

22. Enchhorn.T. Martin.G. - "Course and prognosis in sudden deafness". HNO 1984, Aug. 32 (8) 541-5.

25. Fielder. H - "Clinical study on the use of urografin in the treatment of sudden deafness, necessary preliminary study results and review". HNO. 1984, Aug. 52 (8) 338-40.

24+ Fisch.V.'Managemant of sudden deafness" Otolaryngol. Head. Neck surgery - 1983 Feb. 91 (l) 3-8.

25. Fowler.E.P.- "Sudden deafness" Ann otol Rhinol Laryngol. 59 (1950)

26. Goodhill.V. - "Sudden deafness and round window rupture", Laryngoscope. 61, 1462, 1971.

27. Goodhill (Victor) & Others - "Sudden deafness and labyrinthine window ruptures", Audio - vestibular. Observations, Ann otol Rhinol. Laryngol. Vol.82.1975 (P.2-12)

28. Goto.F. etal - "Hyperbaric oxygen and stellate ganglion blocks for idiopathic sudden hearing loss" Acta. oto - Laryngol. 88 (5-6) 1979, 335-542.

29. Gluckman.J.L. etal - "The aetiology and management of idiopathic sudden hearing loss" S.African Med.J. 50(l) 1976, 15 -18.

50. Greuel.H. - "Suggestion therapy for sudden deafness" HNO 1983 April. 51 (4) 136-9.

51. Gussen - "Sudden deafness of vascular origin. A human temporal bone study". Ann. otol Rhinol. Laryngol. Vol.85, 1976(P.94-100)

52. Gussen.R - "Sudden deafness associated, with bilateral reisseners membrane ruptures". Am.J. Otolaryngol. 1985 Jan-Feb.4(l) 27-52.

55. Hallberg.O.E. - "Sudden deafness of obscure origin". Laryngoscope 66. 1257 (1956)

54. Harbert.F and I.M.Young - "Sudden deafness with complete recovery" Archives of Otolaryngolody. 79, 459 (1964)

35. Hartley.H.V. and Coleman.E.H. - "Deafness of sudden onset. Another case". Journal of Speech & Hearing disorders. 28, 295-299 (1963)

36. Hartley (Harold.V) and others - "Deafness of sudden onset". Journal of Speech & Hearing disorders Vol.29, No.4, 1964(P.482-485)

37. Higgs (Wilson.A) - "Sudden deafness as a presenting symptom of acoustic nauromma". Archives of Otolaryngolody Vol.98, 1975 (P.73-76)

38. Hoke.M - "Sudden loss of cochlear and vestibular function" 1981 (P.198)

39. Hoshino - "Temporal bone findings in a case of sudden deafness and relapsing polychondritis". Acta Oto - laryngolody. 90 (3-4) 1980, 257-261.

40. Hberg.C.V. - "Causal therapy for diseases of the inner ear". HNO 24 (12) 1976, 429 to 430.

41. Igarashi.M. - "Bilateral sudden hearing loss and metastatic pancreatic adenocarcinoma" Archives of Otolaryngolody. 105(4), 1979, 196-199.

42. Ishigani.H. - "Effectiveness of treating by massive doses of ATP, sensorineural deafness of sudden onset." Otol. Tyota 69(7) 1976, 875 to 896.

43. Iwanaga.M. - "Progmostic diagnosis of sudden deafness based on the examination of equilibrium test and electrocochleography." Praot Otol Kyoto 74 (4), 1981, 411-425.

44. Jacobsen.J. - "Sudden deafness" Journal of Laryngology Rhinology Otology. 53 (l) 1974, 822-825.

45. Jaffe.B.F. and Pennar.T.A. - "Sudden deafness associated with hypercoagulation." Trans. Amer. Acad. of Opthal and Otolaryngol. 72 - 774 - 778 Sept - Octo. 1968.

46. Jaffe.B.F. - "Sudden deafness. An otologio emergency". Archives of Otolaryngology, 86, 81 (1967)

47. Jaffe (Burton.F) - "Sudden deafness. A local manifestation of systemic disorders.Fat. Emboli, Hypercoagulation and infections." Laryngoscope Vol.80, No.5, 1970 (P.788-801)

48. Jaffe.B.F. - "Hypercoagulation and other causes of sudden hearing loss." Otolaryngol clin. N. Amer. 8 (2) 1975, S95 - 403.

49. Jaffe (Burton.F) - "Sudden deafness." Archives of Otolaryngology Vol.86, 1967 (P.55-60)

50. Jerger.J.F. etal - "Hearing loss of sudden onset." Archives of Otolaryngology 73, 350 (1961)

51. Kawamoto.H. - "Microwave treatment for sudden deafness." Pract. Otol. Kyoto. 69(8) 1976, 973 to 980.

52. Kerr and Schuknecht (H.F) -"Spiral ganglion in profound deafness". Acta Otolaryngology. Vol.65, 1968 (P.586 - 598)

53. Kikavada.K _ "Three cases of sudden hearing loss". Otolaryngology (Tokyo) 1983 Mar 55 (3) 191-7.

54. Kittamura.K and Komatsozaki. - "Vertigo of delayed onset after sudden deafness and sensory neural deafness followed by mumps". Pract. Otol Kyoto 73 (3) Suppl.2, 1930, 1082 to 1093.

55. Koehn.W, - "Sudden deafness on the age dependance of therapy results with reference to naftidrofulyl". (Dusodril) UNO 1985 Jan. 33 (l) 56-9.

56. Knothe.J. - "Treatment of sudden hearing loss as a clinical problem". UNO, 27(5), 1979, 159-164.

57. Lahnhardt.E. - 'Menieres diseases and sudden deafness". 212(4) 1976, 351-362.

58. Lindsay.J.R. - "Sudden deafness due to virus infection". Archives of Otolaryngology 69, 15 (1959)

59. Lindsay.J.R. and J.J.Zuidema - "Inner Ear deafness of sudden onset". Laryngoscope, 60, 238 (1950)

60. Lumio.J.S. & Aho.J. - "Sudden deafness with special reference to Anticoagulant treatment". Acta laryngology supplement 224, 203,-210 (1967)

61. Lynn (George.E) and Mathswss (Jack) - "Deafness of sudden onset". Journal of Speech and Hearing disorders. Vol.28 No.3, 1963 P.295-296

62. Mair (Iain.U.S) and Elverland (Hans.H) - "Sudden deafness and vaccination". Journal of Laryngology Otology, Vol.91, 1977 P. 323-329.

63. Maurer.R. Karemer.A - "Clinical aspects of sudden deafness". HNO 1904 Aug. 32 (8) 334-7.

64. Mccormick.J.G. etal - "Sudden hearing loss due to diving and its prevention with heparin". Otolaryngol clin. N.Amer.8(2) 1975, 417-430.

65. Mercke.V- "The role of viral infection in sudden deafness". HNO 28 (4), 1380, 125 to 127.

66. Heyerhoff.'.y.L. - "Sudden deafness". Texas. Hod. 72(l) 1976, 80-83.

67. Minami.Y. - "Four case3 of sudden deafness worsen after steroid therapy". Otolargngology 54 (5) 1982, 377 to 383.

68. Mironenkoya.T - "Sudden hearing imprairment and deafness" ORL 58 (5) 1976, 3 to 6.

69. Mizukoshi.K. - "Studies on peripheral vestibular disorders vith special reference to menieres disease, positional vertigo of benign paroxismal type and sudden deafness. Pract otol Kyoto, 75 (Suppl.l) 1982, 172 to 336.

70. Mochio.S. - "A case report on diabetes complicated by bilateral sudden deafness" (Japanese text) Otolaryngol (Tokyo), 51 (12), 1979, 1049 - 1051.

71. Morinitsu.T.etal - "Sudden deafness treated successfully with amidotrizoate"(Japanese text) Otol Fukuoka, 20(5), 1974,599-604-.

72. Nadol (Joseph.B)and others - "Vertigo of delayed onset after sudden deafness" Ann otol Rhinol Laryngol. Vol.84, 1975, P.841-846.

73. Nakamura.K. - "Administration of amidotrizoate in cases of sudden deafness" (Japanese text) Otolaryngol, 47(8), 1975, 521 - 525.

74. Nishida.H. - "Electro cochleographic study of suddan deafness" Otolaryngol 49 (2) 1977, 19 to 30.

75. Ohsaki.K, - "Serum protein in sudden deafness. Study using statistical techniques" otol Fukuokia 26 (6), 1980, 927 to 954-.

76. Oneill.J. - "Sudden unilateral hearing loss attributable to mumps". Journal of Speech & hearing disorders 25, 1960 (55-58)

77. Otto & Kellerhals - "The management of sudden deafness" HNO 24 (4), 1976, 113 to 115.

78. Poneet.E.Etal - "Sudden deafness. Critical study of a series of 118 cases" - Ann.otolaryngol Chir cervicofac, 1983, 100(7) 4-75-8A.

79. Pullen.F.W. - "Sudden hearing loss in divers and fliers." Laryngoscope 89 (9 pt.l) 1979, 1573-1377.

80. Rubin - "Sudden hearing loss". The laryngoscope. 78, 829-833,1968.

81. Saito.H. - "Otosclerosis,cases with postoperative sudden loss of hearing" pract. otol. Kyoto 72 (7) 1379, 871-874.

82. Sakai.H. - "A case of pendreds syndrome with sudden hearing aggravation". Otolaryngol. 53 (5) 1981, 331-536.

83. Saunders.'W.H, & Lippy.W.H. - "Sudden deafness and Bells palsy. A common cause". Ann. otol 68,830-837, 1959.

84. Saunders.W.H, - "Sudden deafness and its several treatments." The Laryngoscope. 82, 1206 - 1213 July 1972.

85. Schiff (Mannice) & Brown (Michael) - "Hormones and sudden deafness". Laryngoscope. Vol.84, No.ll, 1974 (P.1959-1981)

86. Schuknecht (HF) and others - "Pathology of sudden deafness" Acta otol Laryngol. Vol.76, 1973 (P. 75-97)

87. Schuknecht.H.F. - "The pathology of sudden deafness". Laryngoscope 72, 1962,-1142-1157.

88. Schatzle.H - "Treatment for sudden deafness". J.Franc. ORL 30(8) 1981, 495 to 499.

89. Scola.E. - "Sudden deafness". Acta ORL Espan 32(l), 1981, 75-30.

90. Sensini.I. Piccini.A - "Sudden idiopathic hypoacusia. Hematochemical and immuinological investigation." Audiol Fom 1983 Apr. Jun 6 (2) 164-8.

91. Shaia.F.T. etal - "Sudden sensorineural hearing impairment. A report of 1220 cases". Laryngoscope, 86 (3), 1976, 389,398.

92. Shinoxaki.N. - "Clinical statistics of sudden deafness in the last two years". Otol Fukuoka 20(6) 1974, 716-721.

93. Siegel.L.G. - "The treatment of idiopathic sudden sensorirpural hearing loss". Otol Laryngol. Clin.N. Amed. 8(2), 1975, 467-473.

94. Simimons.B.F. - "Theory of membrane breaks in sudden hearing loss". Arch. Otol 88, 41-48, 1968.

95. Singleton (George.T) - "Cervical sympathetic chain block in sudden deafness". Laryngoscope. Vol.81 No.5, 1971 (P.734-736)

96. Stahle.J. and Oberg - "Inner ear deafness of sudden onset as a complication of measles". Nord.Med. 63 (26) 1960, 809-811.

97. Stephens.M.etal - "Sudden hearing loss". Arch.otolaryngol 86 1967, 654-657.

98. Terayama.V.Matashima.J. - "Sudden conductive hearing loss caused by finger manipulation in the ear canal". Archives of Otolaryngology 1985 Jun 109 (6) 425-6.

99. Van Dishoeck. H.A.E. and Bierman.T.A. - "Sudden perceptive deafness and viral infections". Annals of Otolaryngology 66, 963 - 980, 1963.

100. Villar.J. - "Results of treatment of sudden deafness". Acta. CRL. Espath. 32(3), 1981, 3:39 to 331.

101. Wada.J. etal - "A sudden hearing loss". Otal.Fukuoka, 25, (Suppl.l) 1979, 169-173.

102. Weinang.P. - "Spontaneous remission in sudden deafness" UNO 1984 Aug.32 (8) 346-51.

103. Wilmers.H. etal - "Viral infections and sudden perceptive deafness". Laryngology. Rhinology. Otallogy 58(l) 1979. 817-821.

104. Wilson W.R. - Why treat sudden hearing loss". Am. J. Otol.1984 Oct. 5(6) 481-3.

105. Wilson - "The efficacy of steroids in the treatment of idiopathic sudden hearing loss". Archives of Otolaryngology 106(l2) 1980 772 to 776.

106. Wilson.W.R. etal - "Viral and epidermialogic studies of idiopathic sudden hearing loss". Otolaryngol. Head. Neck. Surg. 1983, Dec.9l(6) 653-8.

107. Yagi.T.A. - "Study of the early stages of sudden deafness". Otolaryngol 52 (3) 19 80, 205-211.

108. Yanagita.N. - "Follow up study of sudden deafness". Otolaryngol 53 (4) 1981, 233-238.

109. Yoshimoto.Y. - "Sudden deafness due to acoustic turner". Otolaryngol 53 (9) 1981, 631-639.

110. Zaytoun.G.M. Schuknecht.H.F. - "Fatality following the use of low molecular weight daxtran in the treatment of sudden deafness". Advanced Otorhinolaryngolpar 1985, 31, 240-6.