Sudden Deafness-'A State of Art'

Reg No 10

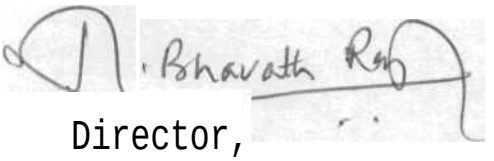
Submitted in part fulfilment for the Degree of M.Sc, Ist year (Speech & Hearing)
All India Institute of Speech & Hearing
Mysore

D E D I C A T E D

T O M Y B E L O V E D P A R E N T S .

C E R T I F I C A T E

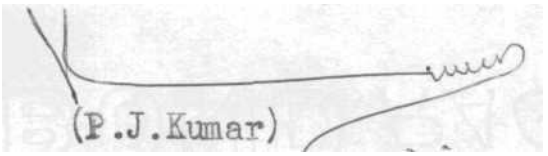
This is to certify that the Project entitled "SUDDEN DEAFNESS" - " A state of Art", is the bonafide work in part fulfilment for the Degree of M.Sc, Ist Year, (Speech & Hearing) of the student with Register No.10



Director,
All India Institute
of Speech & Hearing,
Mysore.

C E R T I F I C A T E

This is to certify that this independent project work has been prepared under my supervision and guidance.



Guide.

D E C L A R A T I O N

This independent project is the result of my own study undertaken under the guidance of Mr. P.J.Kumar, Lecturer in Audiology, All-India Institute of speech & Hearing, Mysore, and has not been submitted earlier at any university for any other diploma or depree.

Mysore,

Dated: Reg.No. 10

A C K N O W L E D G E M E N T S

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CONTENTS

CHAPTER I

INTRODUCTION

SUDDEN DEAFNESS.

The phylogenetically newer portion of the inner ear, which is known as pars inferior, and includes the cochlea and saccule, is relatively more susceptible to various injurious agents than the pars superior. Existing evidence indicates that only a limited number of etiological factors are responsible for acute injury to the pars inferior that results in sudden deafness(Snow '73).

On the other hand, any disease or disorder of the external ear, of the middle ear, of the otic capsule (of the neural connection, also, on occasion, may present with sudden hearing loss. Sudden deafness may be defined as a sensori-neural hearing loss, which develops over a period of hours or a few days. The severity of the loss may vary from mild to total loss of perception of the most intense sound. It may be permanent, of the hearing may spontaneously return to near normal or normal. Although sudden deafness is usually unilateral, it can also be bilateral.

Definitions:

Lindsay('59), Saunders ('67) and Jaffe ('67) define sudden hearing loss as a mild to total sensori neural hearing loss, which is most often unilateral and reaches its maximum within a time range from instanteneous to a few hours on a few days at most.

Goodhill's ('71) view about sudden deafness is "one whose development issudden or instataneous or rapid, i.e., over a period of minutes or hours, which is usually an unilateral loss and is unrelated to middle ear disease or trauma and may be the sequelae of any one of a large number of varying etiological factors".

The same author has given one other definition in '79, according to which the term is used "to describe syndromes of spontaneous sensori neural hearing loss, in patients, with presumably unknown previous ear or hearing problems."

Van Dishoeck ('72) has used the term sudden deafness to conditions in which "hearing loss is the most important or the only symptom developing within

a short time, in a matter of a few minutes or hours, increasing in severity, without any cause apparently".

Simmons ('63) has put forward the idea of sudden deafness as " a symptom in search of diagnosis". Siegel's ('73) definition is "it is an abrupt severe loss of sensori-neural hearing.

Saunders('72) uses the term to describe "a unilateral, generally abrupt loss of hearing andis of type called idiopathic".

Prosper menieres' was the first to report about a case with sudden deafness in 1861. The problem being a manifestation of cerebello pontine angle tumor was first mentioned in 2 cases published by Lushing in 1914. Sudden deafness presenting as the initial symptom of an acoustic neuroma was first described by Hallberg in 1956.

For a long time, the medical people were of the belief that onset of sudden deafness was strictly associated with vascular accidents of the cochlea. This was especially relevant to theelderly people,

who frequently displayed vascular irregularities of one kind or the other. However, as an increasing number of younger people began to display similar clinical manifestations, serious investigation began to determine other possible causes of the problem and today over 200 factors are listed.

Some of these factors as listed by Simmons ('77) are meningitis, encephalitis, sickle cell crisis, mumps with or without, parotitis, multiple sclerosis, syphilis, cogans disease, acoustic neuroma, menieres disease, head injury, co-agulopathics, embolism, metastatic carcinoma and drug toxicities. Besides these, there are still other etiologies coming under the "cause unknown" group. By popular concensus, vital injections rank the highest here, followed by vascular disorders, intra-cochlear membrane breaks and the combination of these causes.

The sudden onset of symptoms is the outstanding feature in this type of deafness. Audiologic findings in them have been highly variable like Jerger('6l) reported of 2 patterns of response in the 12 sudden deafness patients he saw. Herbert & Younp ('64)

observed variation of response, in the same subject, as a function of test procedure, stimulus pattern and instruction. Though different patterns of pure-tone audiograms are encountered, there is a general uniformity. Sudden sensori losses are mainly flat or show a high tone cut off, sensori-neural patterns are almost all high tone and retrocochlear may be low tone, trough shaped, flat or high toned. (Morrison '75).

The hearing loss ias often associated with tinnitus, (appearing at different times, with relation to hearing loss) (Jerger '61), with diplacusis (Jaffe '67), a feeling of fullness or plugging of the ear (Bosatra & De steftani '61). Sometimes vestibular symptoms occur which may be mild to short duration - to days of incapacitation due to nausea and vertigo (Jerper '61). There are few other associated symptoms which although not common, may occur concurrently with the deafness, they are recruitment, pain, fever, upper respiratory infections etc.

Sudden hearing loss has been variously classified. One of the mostly popular classifications is based on etiolopy, i.e., whether it comes under the 'cause known' or 'cause unknown' group. Under the 'cause known' or 'cause unknown" group, (Richards '79): Harbert ('64) has classified the group based on the audiologic test results, into cochlear and neural types, which has been accepted by many other authors like Jerger ('60), Morrison('73) etc. There are also a number of less popular classifications.

The pathogenesis of sudden deafness has been partly understood for most etiologies, except a few conditions like viral disorders, where the understanding has gone a few steps ahead. For conditions like vascular disorders leading to sudden deafness, the explanation is that a thrombosis or an embolism may obstruct some of the most 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 deafness.

But how exactly the different patterns of hearing loss like, sensory, neural or sensorineural is caused, is not explained. These dife-rences have been explained by the viral actiology theory. Viruses can become attached to erythrocytes and cause haemagglutination. They can also cause edema of capillary endothelial ceils and produce a state of hypercoagulation (Jaffe '67). Viruses appear to cause neurological symptoms in two ways: (l) by direct invasion of nervous tissues or (2) by starting some form of antigen-antibody reactions, sometimes the same virus can produce both, leading to a sensorineural pattern of hearing loss (Morrison '73).

The pathology of the end organ caused by the above infections has been studied in detail by Schuknecht in 1962. He reported that the organ of corte was missing in some parts, the hair cell and ganglion cell population was highly decreased. The tectorial membrane, reissners' membrane and stria vascularis were all collapsed, Suceule had also undergone degeneration.

Although a basic identification of the problem of sudden deafness is quiet easy, a definitive diagnosis lies a long way off. A series of steps need to be carefully followed before finally diagnosinp. These steps range right from a careful, general and specific history taking, to a careful examination which again includes general,otologic, audiologic, vestibular and neurologic. Besides these, a series of pathological investigations with X-rays ecochg and more advanced investigations are necessary to diagnose a sudden deafness case.

While doing the above investigations, it is necessary to differentiate sudden deafness from a series of other problems which have a similar symptomomatology like labyrinthine otosclerosis, bacterial labyrinthitis, cerebello pontive angle tumors, trauma, menieres disease etc.

After diagnosis, comes one other very important step, the treatment, which is as controversial as the diagnosis is. Jaffe('61), Meyerhoff('76) etc., are of the view that the occurrence of sudden deafness should be considered as a medical if not a surgical emergency. The sooner the treatment is begun, the better the

results, but as the duration between onset of problem and treatment is increased the prognosis decreases. (Guilton '65). Besides the duration, there are many other factors which are attributable to prognosis, they are age of onset, presence or absence of vertigo, the audiologic Curve, vascular problems etc.

A careful evaluation before beginning therapy is necessary to uncover etiologies, which are potentially dangerous and may be treatable. Specific therapy should be aimed at any specific etiology uncovered during evaluation and non-specific therapy should be utilised to increase cochlear blood flow, reduce intravascular co-agulation and reduce inflam-mation. Approximately, 50% of patients will recover some or all of their auditory function while the remaining 50% will have no recovery at all.

Although a large number of treatment methods exist for sudden deafness, i+ is very difficult to say which is the best, as the frequent spontaneous recovery of hearing to normal or near normal level in these patients make evaluationof any form of therapy very difficult, (Snow '73).

CHAPTER II

ETIOLOGY

Sudden deafness may be the presenting symptom of a wide variety of medical and surgical diseases, such as vascular occlusion, hyper co-agulation, acoustic neuroma, ototoxic drugs, meningitis etc. As a result of the large number of etiologies, there are many competing theories concerning it, and each has righteous follower and indignant antagonists.

The importance of predisposing factors is debatable. Nevertheless, frequently cited predispos-iagfactors include allergic manifestations, changes in physical environment, use of extranel, the emotional state of the patient, fatigue, diabetes, arterios-clerosis, age of patient, pregnancy, and infrequently it is surgery and general anesthesia. The presence of any of these factors and the onset of sudden deafness are most likely chance co-incidence, except in few cases vhere the cause can be pinpointed.

Sudden hearing loss may occur as the result of lesions of external, middle or inner ear or

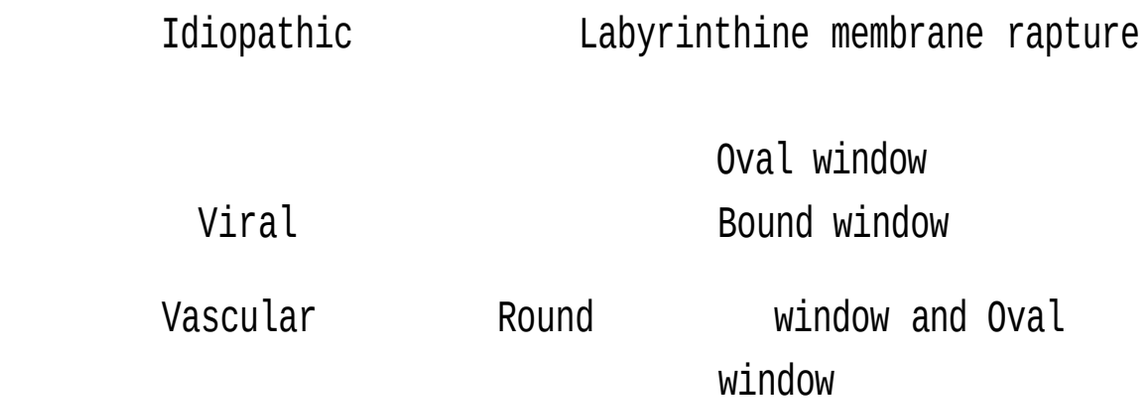
internal auditory meatus, cerebellopontive angle or CNS lesions. The hearing loss may be mild to moderate in degree, more frequently severe or total and may be due to either a conductive or sensori neural lesion. Sudden conductive hearing loss may occur in tympanic or middle ear trauma, in incus necrosis associated with chronic osteomastoiditis, in serous Olitis media following acute upper respiratory tract infection, in post stapedectomy complications and in many other external middle and middle-inner ear interface conditions.

Sudden sensory neural hearing loss may occur in menieres disease, labyrinthine complications of previous ear surgery, acoustic trauma, in response to ototoxic drugs, multiple selerosis, Logan's disease etc.

Sudden mixed hearing loss may occur as a result of labyrinthine fistulas, in chronic ostomastoidities, after stapedectomy and in other complications of otologic diseases.

Until recently all cases of sudden deafness were considered to be of idiopathic etiology, as the research was not sufficient to identify the basic etiopathologic cause, although, presently some of the hypothetic and speculative approaches have made possible

an accurate diagnosis in some cases. The other problems in diagnosis are the presence of limited literature on validated temporal bone pathologic changes and the high occurrence of spontaneous recovery in these cases. Goodhill ('79)[167] has indicated the etiology of sudden hearing loss, for an easy understanding by the following figure.



Rchiff & Michael('74)[17] have recorded the following possible causes:

I- Infection.
   (a) Viral (b) Bacterial
II. Traumatic
   (a) head injury (b) rupture of oval window/round window.

III .Tumor formation.

(a) acoustic nearoma (b) Vascular malformations.

IV.Post Operative.

(a) Ear surgery (b) complications from other far distant surgery.

V. Pregnancy.

VI. Endolymphatic hydrops.

VII.Metabolic.

(a) Ototoxic drugs (b) disturbance of lipid metabolism.

VIII.Vascular.

(a) associated with hypertension and/or diabetes
(b) arteriosclerosis
(c) alterations in blood co-agulation
(d) vasculitis inducted or immunologic.

Blair Simmons ('68)[148] has also recorded similar etiologies.

Besides the above given etiological factors there have also been reports of some miscellaneous rarities like Gopichand ('68)[132] reported of a case who developed sudden deafness after a severe emotional excitement. A search of the literature would

undoubtedly reveal many other rare causes of sudden hearing loss.

There have been reports of the incidence of the problem with respect to specific etiologies. Some of these are by Jaffe & Arbor ('67) [123] Morrison ('71) and Morrison ('75) [122]. Of these, the one reported by Morrison in ('75) is the most extensive one and is as given in the table.No.l. The above listed causes will, inthe following paragraphs, be discussed in detail, which according to the classification given by Anthony ('76) [152] fall into the category of 'Known' causes of sudden deafness.

Central Causes

1. Diffuse cortical eucephalitis:- It is presumed to have a viral origin which affects the substance of brain. It can cause a wide variety of problems of which sudden hearing loss is one. It may also cause auditory aphasia when both the temporal lobes are involved. The condition is confirmed by EEG evaluation. The patient will have normal puretone audiogram, yet no understanding of speech. The illness develops slowly over weeks or months with fluctuating symptoms and acute episodes. Features include headache, drowsiness, personality changes, generalised epilepsy,

aphasia, mutism, sudden deafness, parasthesia and abnormal movements. In the beginning stages it is usually confused with psychogenic deafness. Euce-phalitis as a cause has been put forward by Schiff et all ('74)[165] Simmons ('68)[176] & Morrison ('71)[123].

2.Psychogenic deafness:- It is not an uncommon condition and has many reasons for its occurrence,e.g., compensation. The impression of psychogenic deafness is aroused by the clinical impression and by incon-sistencies in the story and the findings. Tinnitus is seldom a feature. The different tests like lambard, stenger etc., can be used to identify the condition. Morrison has reported it in both of his studies i.e., of 1971 and 1975.

Retro cochlear causes:-

1. Acoustic Neuroma:- Sudden hearing loss, presenting as an initial symptom of acoustic neuroma was initially reported by Hallberg in 1956. Wilson ('73)[195] is of the view that sudden deafness is an unexpected symptom and found its occurrence in 10% of his cases. Nedzelski ('75)[138] and Morrison('75)[122] have also reported similar findings. More recently the condition was surgically confirmed by Kusak ('79)[100] in some of his cases with sudden deafness.

TABLE No. 122

Given by Morrison('75). Cause and Ape group in 218 patients.

Diagnosis	No. of cases	Unilateral		Bilateral		Mean age
		Male	Female	Male	Female	
						years
CENTRAL						
Diffuse Euce-phalitis	3				3	30
Sudden Psycho-genic	12	3	1	2	6	18
Ratio Cochlear						
Acoustic Neuroma	14	6	8			42
Multiple Sele-rosis	7		3	2	2	33
Lepto Meningitis	8			8		27
T.B .Meningitis	2			2		21
Syphilitic Meningitis	3	1		2		41
Meningo Myelitis	2		1		1	24
Bacterial Endo-carditis			1			69
Cochlear						
Late syphilis	13		2	5	3	41
Memeres disease	17		4	3	4	38

Diagnosis	No.of cases	Unilateral		Bilateral		Mean age
		Male	Female	Male	Female	
						years
Cochlear Oto-selerosis	3		1	1	1	44
Viral Zoster	7	4	3			42
Mumps	14	6	8			12
Measles	4	1	2		1	8
Suppurative labyrinthitis	6	4	2			45
Vascular Lesions	15	9	4	1	2	64
Rarities	7	3	3	1		34
Idiopathic	59	32	25		2	36
Trauma						
Fracture Otic Capsule	5	4	1			28
Stapes facture	8	5	3			26
Blast sensory loss	4	1	1		2	29
Decompression deafness	4	4				27

Cochlear causes:-

1. Syphilis:- Whether congenital or acquired, syphilis is still an important cause of sudden deafness. The incidence of sudden deafness in various forms of syphilis as reported by Joseph ('75)[127] are:

Late congenital syphilis	-	18%
Early latent "	-	17%
Late latent "	-	25%
Asymptomatic neuro "	-	25%
symptomatic neurom "	-	80%

Both the early and late forms of congenital syphilis as the cause have been recorded by Lloyd ('78) and on the other hand Thomas ('78) has sudden deafness in both secondary and tertiary forms of acquired syphilis. The clinical picture in both forms are similar having sensory neural loss with low discrimination scores affecting both ears simultaneously or sequentially vestibular symptoms are a common occurrence here. Morrison ('75)[122] Joseph ('75)[127] and Nelson et al ('74)[132] are the other researchers who have reported syphilis as a cause of sudden deafness.

2.Menisres disease:- Sudden unilateral sensory deafness and tinnitus herald the onset of idopathic endolymphatic hydrops in about 3 percent of patients with this disease (Morrison '75)[122]. Once the full clinical picture of hydrops has developed the diagnosis is obvious. In the early stages it is inferred by audiological features of hydrops and by significant change following hydiops. Sudden deafness may sometimes affect the second ear after an interval of months or years. Morrison ('75)[122] The topic is discussed in more detail under the Chapter of differential diagnosis.

3.Cochlear Otoselerosis:- Evidence of cochlear involvement in otoselerosis is very common though sudden spontaneous total or sub-total deafness in cochlear otoselerosis is a very rare event. Morrison ('75)[122] has given the most likely explanation for the occurrence of the above two. He says that there may be rupture of the cochlear adjacent to an area of endostead disease and in stapedectomy also the spontaneous rupture of cochlear duct can be expected with sudden deafness is the result. It may arise from a viral or vascular pathogenesis, but in the absence of such evidence it can be due o surgery

or as a hazard of stapedectomy. But whether this loss is due to other reasons which coincided, is not well known as in some cases sudden deafness has been reported to occur in unoperated ears. Armstrong ('67)[3]. Sale ('69) & Morrison ('75) are the others who have also reported the occurrence of sudden deafness in otoselerosis patients.

Ototoxic drugs:

A large and increasing number of drugs are now known to be ototoxic. They usually cause a progressive sensory deafness though in certain conditions it might be sudden, bilateral, severe and continuing even after withdrawal of the offendingmedication. In the presence ofrenal failure rapid accumulation of the drugs and high concentrations in the endolymph can result in alarmingly rapid sensorineural deafness. Morrison ('75)[122]. The different ototoxic drugs are kanamycin,nevamycin,streptomycin, ethacrynic acid, diuretic agents, saliculates, quinineand furosomide. Kanamycin and neomycin are supposed to alter cochlear function while gentamycin and streptomycin cause injury to the vestibular system. Of these, the effect of Kanamycin has been reported to be the greatest, as here, there damage of hair cells which is followed by neural damage. The damage of hair cell starts in the basal coil and progresses in the apical direction.

Topical ototexicity has also been found to play a role in sudden deafness, as it has been found that preparations containing neoxycin, polymycin B, gentamycin and framycetin are used in patients with chronic perforations of the drum. Here there is a possibility of penetration of the drug through round window membrane which in turn damages the inner ear leading to sudden deafness. Ballantyne ('70)[8] mentions a severe sensorineural deafness which may have resulted from local framycetin in a patient with chronicotitis media. Similar finding has been reported by Taylor ('75)[189].

Acoustic trauma:

In addition to noise-induced deafness, which can be divided into a slowly progressive deafness caused by prolonged intense noise exposure and acute acoustic trauma caused by a single exposure to very intense sound, Kawata & Suga ('67)[188] & David ('80)[26] have described a noise induced deafness which occurs suddenly after a certain period of exposure to uniform, intesnve noise to which they have long been accustomed. Boenninphams ('62)[14] is of the view that insome cases sudden deafness occurs while physically strenuous labour is carried out inthe presence of noise. Maures ('62)[113], Robert('69)[154]

have also reported of cases in whom the above relation was observed.

Head Injury:-

A head injury with or without the temporal bone fracture can lead to sudden deafness. A concussion type injury with anoxia and or haemorrhage can be the etiologies, as isolated or separate entities. In part the magnitude of the initiating force and patient's level of concieusness can be used as a guide to the potential of sensorineural deafness.

It has been experimentally shown by Richerds ('78)[152] that a total vascular occlusion and associated anoxia can leadto damage of hair cell and stria vascularis.

With a transverse fracture of temporal bone on sensorineural type of hearing loss is usually found and a conductive type of hearing loss with longitudinal fracture: Piaget('64)[146] has reported of cases in whom the above condition was observed.

Miscellaneous Rarities:-

Besides the above given causes, there are some conditions which have on occasion led to sudden deafness

Morrison ('75)[122] reported of a case in whom typhoid fever was followed by sudden deafness. Total unilateral deafness following nitrous oxide inhalation has been reported by Mack ('76)[144] &Morrison('75)[122] Cardio-pulmonary surgery as a cause was put forward by Saunders('72)[162]. Tumours like cholestealoma, of glamus jugulare (Morrison'75)[122] , bronchogenic carcinoma (Vonne '78)[10] were uncovered as etiologies. Re-vaccination has been reported by Klttel('64)[97] Iain('77)[110] and Wirth ('73)[198], Moshe ('73) brought out insecticide poisoning. Gaillard ('66)[47] electric shock and Nakagawaki ('77)[129] shoulder pain, in this broad category of etiology.

A search of literature will undoubtedly reveal many other rare causes of sudden hearing loss.

After having finished the category of 'Known' causes of sudden deafness we can now go on to the 'Unknown' causes which can alSo be called as the 'Mechanism of sudden deafness". This group has been broadly classified under three theories, namely, (1) Viral theory (2) Vascular theory and (3) the membrane rupture theory.

Viral Theory:-

This theory like others has many many righteous followers and indignant antagonists. The protagonists state that in any large series of patients with sudden deafness about one in three has an overt respiratory infection at the time of onset of sudden hearing loss indicating a true relationship. Histopatholopical studies have shown parallel changes to be there in congenital viral deafness and acquired idiopathic sudden deafness.(Jaffe). The antagonists counter with the facts that it is highly improbable that the infection will involve only one ear specifically and secondly the infection often clears whereas the deafness persists in many cases.

Inspite of the controversies the theory has gained support from many researchers like Bull & Ransome ('76), Jaffe ('67)& '78, Malik ('74),Shaia & Sheeny ('76), Van Dishoeek ('74), Morrison('75),Snow('73) and Rowson ('73).

The sites of viral induced pathological changes are the cochlear and the components of 8th cranial nerve. (Snow '73). Some of the virus have also the

capability of affecting the vestibular auditory end organ (Goodwill '79). When the cell injury does not proceed to cell death, a broad spectrum of recovery pattern is seen. Audiometrically, a profound sensori-neural loss is the most common pattern seen in these cases. The magnitude and configuration of the hearing loss, initially, as correlated with that at the time of onset, has been used as a prognostic indicator for return of function. As a rule the more profound the hearing loss, initially, the less the chance of complete recovery (Goodwill '79).

The mechanism involved in the causation of sudden deafness by the viruses has been variously explained. Williams (&'78) has given the following explanation:

(1) neuronitis or ganglionitis / a viral infiltration of the ganglion or nerve, especially seenwith herpes Zoster.

(2) Endolymphatic labyrinthitis(invasion of cochlear duct by the virus.

(3) Perilymphatic labyrinthitis(invasion of the perilymphatic space by a viral agent.

(4) Vascular occlusion (by viruses).

Jaffe & Arbor ('67) are of the view that there are three mechanisms by which the virusparticles can effect blood flow through capillary network of the inner ear and in turn producing sudden deafness.

(1) Virus particles can attach to the erythrocytes and cause hemaglutination in viro.

(2) Virus can affect blood flow by causing edema of the endothelial cells of the capillaries.

(3) Viral infection may produce a hypercoagulable state.

The list of viruses associated with sudden deafness as put foward by Jaffe ('78) are:

l.Upper respiratory infection		1. Adeno virus
2.Mumps		2.Mycoplasma
3.Measles		3.Parainfluenza (type 3)
4.Herpes Zoster	4.Herpes	homines

A	B
5. Chicken pox	S.Herpes Zoster
6. Infectious mononucleosis	6.Mumps
7. Herman measles	
8. Polio virus	
9. Columa&K.Virus	
10. Yellowfever virus.	

Column A lists the viral clinical syndromes associated with sudden deafness and oolumn B are the viruses documented by laboratory tests.

1. **Upper respiratory infection**:-

It forms the "catch all" category which includes the largest viral group associated with sudden hearing loss (Jaffe '78). He has also given the incidence of upper respiratory in patients with sudden deafness.

Year	Author	No. of patients	No. C URI	Percentage of patients' C-URI
1963	Van Dishoeck & Burman	150	50	33%
1967	Jaffa	143	28	20%
1975	Rawson	39	10	25%
1976	Shaia Sheehy	1220	100	8%
1977	Byl	32	7	

As the symptom complex for a wide range of viral infections is same, it is important that a careful virologic work up is done for a differential diagnosis under this category (Jaffe '78). Authors like Sataloff & Vassallo ('68), Snow ('73), Beal('67) Schuknecht ('73), Barton ('71) have all come up with either clinical or experiemental evidence to support the above co-occurrence. But it isstill not clear whether occurrence of upper respiratory infection and sudden deafness is a chance co-incidence or if there really exists a cause and effect relationship between the two (Jaffe '78).

2. Mumps:

Historically mumps was the earliest virus suspected of causing sudden deafness. Here the deafness usually occurs a few days to two weeks after the onset of parolitis,although Daves ('63) reported of a case in whom deafness developed 3 days before the parotitis. Toyubee (1860) was the first to report of sudden deafness in mumps.

Three different forms of hearing loss is found to be concurrent to mumps (Morrison'75):

(1) the unilateral total deafness (2) partial deafness occurring mostly inchildren (3) severe bilateral deafness.

Williams ('63) is of the view that adolescents are more likely to be affected by this condition. John('60) reported of a definite audiometric configuration here which was characterised by B C ths being characteristically lower than the A C ths, with the speech reception ths.being higher than the average puretone loss;discrimination scores were low; there were also indications of recruitment.

The other authors who have come up with evidence of the occurrence of sudden deafness in mumps are Evenberg('57), Lindsay ('60), Saunders & Lippy('59), Richards ('78), Danilides('77), Jaffe ('78),Williams('63) Reynier ('64), Welsh ('63), Prasad ('63). Rawson ('75).

Reynier ('64) and Welsh ('63) have also reported of the occurrence of vestibular symptoms along with sudden deafness.

Measles:-

Clinically cases of measoles having sudden deafness are uncommon, but have been described. Lindsay & Memenway ('54) were the one to report of pathological changes in these cases which also formed one of the

earliest findings to support the viral theory of sudden deafness. Morrison ('75) has also reported of sudden deafness in measles.

Herpes Zoster: (Hunt's syndrome).

Although Herpes Zoster usually involves the outer ear with painful vesicular eruptions, it Ray be associated with facial paralysis and even more rarely with sensori neural hearing loss.

Adeno Virus Infection:-

The association of adeno virus infection to sudden hearing loss was first found by Jaffe & Massab ('67) who demonstrated its role by isolation of the virus and by demonstrating a diagnostic rise in antibody titer to the same virus. Jaffe confirmed this in his study in 1970. In his earlier study he also found an association between sudden deafness and virus in chicken pox. Gregg & Shaffer ('64) included infections mononucleoisis ia the etiology.

Some of the other viral agents that have been associated with sudden deafness are German measles (Jaffe ('78), Polio virus (Mawson '63) Columbia SK

virus (Van Dishoeck et al '57), Yellow fever virus (Moe '47), Eaton Agent infection (Van Dishoeck '63), rabies infection (Jaffe '67), Cytomegalo viral infection (Mitiske '78).

Besides clinical and histopatholopical evidence to support the viral theory, experimental evidence has been provided by Jaffe & Maasab ('67,'73), Van Dishoeck '63) & Rawson et al ('75).

The mechanism by which viruses cause sudden deafness has still not been explained. One might speculate that the virus may directly involve the inner ear by travelling via the blood to the stria vascularis, into the endolymph and then on to the hair cells, via the cerebro spinal fluid to the peri-lymph fluid or via the eustachoan tube to the middle ear and across the sound window membrane to the hair cells (Jaffe '78).

Jaffe '78 has also speculated an indirect involvement such as a virus triggering intra-vascular co-agulopathy with clotting in the stria vascularis or spiral vessels and affecting the function of the inner ear. The viruses may trigger

a biochemical alteration of the endolymph affecting the hair cell but allowing a reversible hearing loss.

Although a cause and effect relationship has not been proven, a frequent association between viral infection and sudden deafness allows a strong argument in favour of viral etiology of sudden deafness.

Vascular Theory:

This theory assumes that thrombosis or embolism occur in small end arteries supplying the cochlea and vestibular labyrinth or both, resulting in sudden hearing loss. Earlier it was thought that this can occur only in older people who present one or the other type of cardiovascular or circulatory problem. But recently it has been found that the same can happen in younger individuals, where the viruses can attack R B C to produce subsequent hemagglutination, causing a swelling of the endothelial cells lining capillary walls, which finally leads to hyper co-agulation. All these factors cause small vessel thrombosis (Saunders '72).

The different conditions leading to sudden deafness of vascular origin are vaso spasms, thrombosis,

embolism, inferection haemorrhage, hyper ooagulation and aiudging of blood (Snow '73) Paola '67). Sudden deafness in this condition may be a localised form of systemic disorder. (Jaffe '70). There is a sudden loss of vestibular and auditory functions and prognosis is reported to be not really good. The abruptness of loss is a reminiscent of a catastrophy (Schenions '77). Such severe and permanent loss is a for the reasons that cochlear tolerance for ischemia is very limited and the Cochlear microphonics and action potential almost disappear after 60 sec. of anoxia, and the potentials remain depressed for a long time even after normal blood flow is restrained.

Reversible loss has been associated with sickle cell crisis, fat embolism, vaso spasm, blood sludging (Sheeny, Towler, Jaffe). The other authors who have given clinical evidence to support the theory are Ruben ('68), Skovrousky ('77) Giaccai ('65), Mironeuka ('76), Harbert & Young ('64), Saunders &3 Lippy ('59).

Vasospasm has been attributed to stress fatigue, emotional state of patient and allergic reactions of the antigen-antibody type. The hearing loss occurring

fromthis is characterised by severe hypacusis of cochlear origin with recruitment (Paola '67). He also observed that the serum aldolases would increase in the days immediately following the onset of symptomatology and returning to normal value after 10-20 days which he attributed as the possible reason for spontaneous recovery. Snow ('73), Gopichand ('68) & Wedaver ('73) have all reported cases in whom vasospasm was the cause of sudden deafness.

Thrombosis and embolism are usually attributed to arteriosclerosis and hence is more frequent in older people. The effects that a thrombosis can have are cited by Paola ('67) as (1) thrombosis effects the labyrinthine artery or the internal auditory artery immediately after its origin, causing a complete suppression of the function of both the anterior and posterior labyrinth (2) thrombosis of the common cochlear artery causes, a complete deafness, here the function of the posterior labyrinth supplied by the vestibular artery will not be affected. (3) and the thrombosis of the cochlear artery proper, there will be deafness which will be more or less severe but not total, as a great area of the cochlear will be still

supplied by the cochleo vestibular artery. Jaffe ('67 & '70) has also reported similar findings.

Although embolism formation is postulated as an etiology of sudden deafness, there is not much evidence to accept or reject it. But Jaffe('70) Dubs ('56) and Snow ('73) are of the view that it does contribute in occurrence of sudden deafness.

Clinical observations in the form of microscopic studies of fingernail beds, clumping of red cells in retinal vessels and dumping of red cells in vitro have given rise to suspect that sludging of blood can also give rise to sudden deafness (Towler '57). For this condition, clinical evidence has been given by Ohsaki et al ('75).

Sickle cell anemia as a cause has been put forward by Morgenstein & Menace( ), Sergent et al ( ), Daniel ('77), Urban ('73).

There are several reports of sudden deafness associated with leukemia, haemorrhage into the cochlear is considered to be the cause of the loss.

Clinical evidence for this has beenprovided by Kock ( ), Schuknecht, Sando ('77), Igarashi ('65), Keleman ( ).

The less known vascular abnormalities leading to sudden hearing loss are (1) Aneurism of basilar artery (Arnold '77), (2) polycythemia Vera (Magni '68 & Davis '65). (3) Labyrinthi apoplexy(Ritler'77) (A) Thromboaigitis obliterans (Kirikal '62), (5) diabetes Mcllitis (Jorgeusen '66) Akelson& Jayerberg '68).

Experimental evidence for the vascular involvement in sudden deafness has been provided by Snow('73) Kiniura & Pearlman ('56 & '58) & Ruth ('77).

Membrane Rupture Theory:

Simmons ('68) was one of the authors who initiated the fact that pressure changes in the inner ear due to sneezing, coughing and other physical activities like lifting, diving etc, tend to raise intra thorack, intracranial or C.S.F. pressures which would break or rupture the membranous cochlea, which could in turn give rise to sudden deafness.

Ahlea showed a correlation between peri-lymphatic pressure and respiration; abdominal pressure and compression of cervical blood vessels. Labyrinth is a delicately balanced perilymph-endolymph system which is exquisitely compartmented by delicate membranes. So any sudden pressure changes in these areas can rupture the labyrinth. (Goodwill '71).

Sean ('77) is of the view that sudden increase in intracranial pressure or middle ear pressure may leak through the cochlear aqueduct, into the inner ear and in turn damage it. These perilymphatic fistulas also form a serious and common complication of stapedectomy (George et al '). In some cases there may be 2 membrane breaks, one at the oval or round window and the other further inside the cochlear. Such a condition is labelled as 'Double-membrane Break Syndrome' (Simmons '79).

Farrior & Endicotl ( ) & Goodwill '72 are of the view that a patent cochleo or an abnormal cochlear aqueduct exists in these patients, which makes it possible for the increased C.S.F. pressure to be transmitted to the labyrinth.

Sudden hearing loss due to perilymphatic fistula has also been reported to occur in children. (Kenneth '78)

Goodwill ('71) has put forth two basic mechanisms of formation of the perilymphfistula i.e., through explosive and implosive membrane rupture routes.

1. Explosive route:-

According to this theory, when there is an exertion, there is an increase in C.S.F. pressure. This pressure is transmitted to perilymph system either through oochlear aqueduct or internal auditory meatus, of which the former is supposed to be a more logical route.

The author has also suggested that an infantile type of cochlear aqueduct may persist in some adults, with a relatively large cross-sectionsl area, thus losing its protective effect. On the other hand it more easily passes on the C.S.F, pressue to the labyrinthine areas. The laminae cribrosae are also potential channels for the transmission of pressure. Thus cochlear aqueduct and

possibly the I.A.M. route play arole in explosive apertures.

2. Implosive route:-

This occurs in valsalive maneuvres, where the sudden air pressure increase is transmitted via eustachian tube and the middle ear, on to the inner ear, thus resulting in a sharp increase of intra-lymphanic pressure, which may rupture either the oval window annular ligament or the round window membrane.

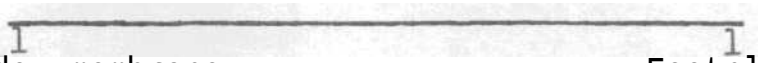
EXPLOSIVE ROUTE

	C.S.I.	C.S.I.
	Cochlear aqueduct	Internal acoustic meatus
Round Window ligament	Scala tympam	Lamina cribosa
Middle ear	Basilar Membrane	Saccule & utricle
	Scala media	Vestibule
	Reissners Membrane	Footplate Ligament
Footplate ligament	Scala vestibuli	Middle ear
Middle ear	Vestibule	
	Saccule & Utricle	S.C.C.
	Footplate ligament	
	Middle ear	

IMPLOSIVE ROUTE

Valsalva - Eustachian Tube

Middle ear



Round Window membrane		Footplate
Scala Tympani	Scala	Vestibule
etc.		etc

Pullen ('79) is of the idea that the susceptibility of the rupture is related to the angle of the round window membrane, whereas Paul('78) says that the more the surface area the greater the chances of rupture. Weakness of the anular ligament of the stapes footplate or round window membrane coupled with a rapid equilibrium system was found in the above condition (Strand '79, Goodwill '72).

In most cases there is a history of physical exertion, vertigo, tinnitus with sudden deafness. A large percentage of the cases shows spontaneous recovery (Goodwill '72).

The clinical and surgical explorations put forward to support the above theory were by Fee('68), Farrior& Endicote ('72), Grace ('76), Nedzelski('76), Terge ('78), Edmonds ('73), Farmer ('76), Freeman('74), Goodwill ('71) and '72 & '73),Healy ('74), Pullen ('72), Steoud & Calcaterea ('70), Healy ( ), Kenneth('78).

Divers have been most frequently associated with this theory. Inner ear Sarotrauma, decompression sickness, gas embolism, a change of breathing gas during leading to counter diffusion - are the most

frequently postulated causes in divers (Molifer '79) Commander ('79), Molvfer('79), Freeman & Edmond '78) have also given clinical reports of the occurrence of sudden deafness indivers.

Clinical and experiemental evidence has revealed that rupture of round window membrane by itself is not a major cause of sensorineural deafness. Even the sites of the primary trauma have not yet been discovered, but it would seem reasonable in the light of the recent reports that the perilymphatic vessels, the stria vascularis and/or the integrity of the coehlear duct might be involved in the genesis of the hearing loss (Ales et al '78). They have also suggested the use of the term 'Round window rupture syndrome' to be used in the above condition.

CHAPTER III

CLASSIFICATION.

There are very few classifications that have been provided by different people in order to categorise the group of sudden hearing loss. These classifications are from different points of view i.e., some of them totally concentrate on the point of etiology whereas others go beyond it to classify depending on the degree of loss, the type of loss (i.e., cochlear or retrocochlear) etc. Each of them have their own limitations, in that a classification of etiology does not bother about the degree of loss and vice versa. As a result of which a classification, so chosen, depends upon the orientation.

One of the broadest classifications is the one given by Simmons( ). This is based on etiology and divides the group into 2 main categories: (l) Cause 'known'. (2) cause 'Unknown'. Bolognesi ('60) has said that a better understanding could be achieved by dividing the group into 3 sub-divisions, namely (1) those cases with an

immediate obvious cause (2) those with a cause which becomes obvious and (3) those which remain of obscure etiology. such classifications have been accepted by many people who have also provided further sub-division, to the major categories. This topic has been dealt with in great detail under the Chapter of 'Etiology'.

Rubin (68)[144] has provided a classification based on the severity and audiometric configuration of the hearing loss. In type I, according to his scheme, the hearing loss is mainly in the lower frequencies, with some elevation of threshold from 2 to 8 kH_2 The speech reception threshold is decreased and the discrimination is severely depressed.

Type II includes more uniform and severe elevationof thresholds for puretones, with a 50 to 60 db loss for the speech frequencies i.e., 500KH_2, 1 KH_2 and 2KH_2 There is a sharp loss above 3KH_2. The speech reception and discrimination scores are consistent with what would be expected with similar puretone thresholds in more gradually developing forms of hearing loss.

Type III is a complete loss of hearing with no measurable discrimination.

Sheehy ('60)[190] classified sudden deafness into 4 groups; low tone, flat, high-tone and total hearing loss. A flat type of hearing loss was found in 41% of his patients. The next most common type was the high-tone loss, and this type accounted for 29%. The low tone type made up 17% of the group, and total loss of hearing occurred in 13%.

A'sudden hearing loss' research clinic was set up at Oakland in '73 which had 30 members of WNT who referred cases of SHL. This team was headed by Frederick, they saw sudden deafness cases and classified the group as having mild, moderate or severe loss, according to the below column. Here average of speech frequencies was taken for classi-

Mild	34 dB
Moderate	35 - 54 dB
Severe	55 - 74 dB
Profound	75 dB

Rased on the audiological findings of SHL cases, they have been divided into 2 groups cochlear and neural (Harbert '64) & Myran ('67). In the first group there was no significant tone decay and separation between pulsed and steady tone in Hekesy was absent or minimal. These patients often complained of fullness in ear, diplacusis, hypercuse and distortion. The second group showed abnormal adaptation, wide separation between pulsed and steady tone in Rekesy. There was absence of recruitment and discrimination was poor.

Such a classification has been reported by many other authors. Jerger ('61)[143] tested patients with unilateral sudden deafness and found 2 groups existing in them. The first group had the following characteristic findings:-

Type II Bekesy tracing, SISI scores from 60 to 100%, slight recruitment on ABLB and speech discrimination scores from 2% to 46%. The second group had characteristics like Type III Bekesy, SIS1 scores were from 25% to 40% absence of recruitment and none respondent to speech discrimination. Hence he concluded that the group could be divided into cochlear and neural respectively.

Electrocochleo graphic studies have also been done to arrive at the above classification. (Graham et al. Hiroaski et al & '78). Ecochg was performed using techat ques given by Aran & Lebert '65 & by Yoshie '68. 3 Types of electric potentials have been described:

Action potentials:

(1) AGP representing 8th nerve activity.

(2) Cochlear neurophonics recorded from the outer hair cells close to round window membrane and

(3) Summating potentials which gives an indication of basilar membrane displacement.

To distinguish between cochlear and retro-cochlear the following criteria was used:-

1. Cochlear:

1.Diphasic action potential (commonly associated with hair cell loss.

2.Widened action potential complexes with multiple or double peaks (seen in mehieres disease, syphilis and other cases of endolymphatic hydrops.

3. Input/output latency function showing recruitment.
4. Presence of an enhanced summating potential indicating endolymphatic hydrops.

2. Retro-cochlear:-

1. Widened action potential often with large cochlear microphonics.
2. Definite cochlear microphonics in absence of action potential.
3. Action potential threshold better than subjective hearing threshold. Speech audiometry has also been found to be of use in differentiating above 2 groups. (Jaffe')[168].

Perilymph fistula has long been considered as one of the major etiologic factor of sudden deafness. Kenneth ('78) has piven a classification of the fistula and accordingly the sudden deafness resulting from it. It is as follows:-

Congenital:

Communication between inner ear and middle ear is present at birth without associated temporal bone or extracranial abnormalities e.g., isolated stapes footplate fistula.

Acquired: Normal ear anatomy at birth.
Ratrogenic Post-stapedectomy.

Traumatic:

Direct penetrating wound in the ears
Indirect blunt injury to head

Barotrauma:

Implosive alternation of tubotympanic pressure
Explosive alterations in C.S.F. pressure.

Erosion of bone:

Luetic
Cholestealoma
Neoplasm.

Combined Underlying anatomic abnormality that makes the ear susceptible to injury from alterations in pressure in perilymphatic fluid or middle ear or both.

Theclassifications of Rubin and of Sheehy are useful primarily because they are simple and easy to keep in mind. Of equal importance to the severity of the hearing loss and its audiometric configuration is the presence or absence of associated vestibular symptoms and signs. Likewise, the

rapidity of onset of the hearing loss is important, but a classification including all of these factors becomes so elaborate that its usefulness is limited. (Snow '73)[173].

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CHAPTER IV

Pathogenesis and Histopathology

Pathopenesis:

There are 3 routes of access of viral particles to the inner ear. The most common is a viremia, in which the virus are deposited within the membranous cochlea. The second route is where virus 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) & Snow'(73).

Transmission of viral infection from brain to labyrinth has also been reported (Erich ('76). Here there is an infiltration by lymphocytes and histocytes along the nerves in the internal meatus into the spiral ganglia and the scala tympani (Lindsay '59). There is an infiltration of various cells like histocytes, lymphocytes, plasma cells into the inner ear which leads to fibrosis and then a disappearance of some sensory nerve cells. (Blackley '7)

When the viruses become attached to erythrocytes and cause haemagglutination and edema of capillary endothelial cells which in turn produces hyper co-agulation, a sensory loss is seen(Jaffe '67). But when they invade the nervous tissue or start some form of antigen -antibody reaction a neural loss is seen. Sometimes the same virus can produce both (Morrison '70). The relation between direct neural damage and edema from antigen -antibody reaction decides the extent of recovery (Morrison '70).

Once the virus particles gainaccess to the membranous cochlea, pathophysiological changes start occurring which are often reversible, but once the destruction is extreme, the loss is permanent. Initially the vascular endothelium is invaded and inclusion bodies may be seen in the endothelial cells. Here the capillary endothelium is often swollen and may proliferate as lumen is narrowed, there may be reduction of blood supply. But frequently there is hyperemia at the site of inflammation which prevents this from happening. Hemagglutination and sludging of blood

occurs when virus particles attach to erythrocytes. When there is inflitration of different cells edema in perivascular spaces occurs. This may lead to necrosis of neuroepithelium. (Snow '73).

Morrison('75), Jaffe '67), Welsh & Welsh('63) Gregg & Shaeffer ('64) have all been unable to isolate viruses of any type intheir viral antibody studies, C.S.F. studeis, stools or postnatal swab studies or by the Paul-Bunnell test, in their clinical studies.

Alteration of the immune response in the grand state, antigen-antibody reaction, or hypersensiturty have all been reported as the cause of suddendeafness,in conditions where pregnancy has lead to suddendeafness (Morrisont'73; Jaffe ('67) Welsh & Welsh ('63). Morrison ('73) has also reported a high occurrence of false positive, biological test results like VDRL, CWR or RPCET, which may be temporary or chronic.

As mentioned earlier otiticbarotrauma is a frequent cause of perilymphatic fistula. Kenneth('73)

has put forward his explanation for the. He says that the eustachian tube is lined by a surface active membrane (3AM) which has surface tension lowering properties similar to pulmonary surface-ant. It allows Sheilamenal wall of the tube to separate in response to the pull of the muscula-ture. It gets destroyed by proteolytic enzyme producing bacteria or in smokers. This in turn affects the pressure equalising capacity of the tube. It is in these conditions that otitic baro-trauma occurs. Cochlea may also be injured when there is abnormal air pressure build up in eust-achian tube or middle ear which results in an inward and outward displacement of the stapes footplate in the oval window, creating pressure distortion of the membranous labyrinth (Berkley'70) In these conditions besides pressure, haemmorrhage from torn vessels and gas bubbles released, can also damage the cochlea. (Stircker& ESchob).

Perlymphatic fistula may also result from rapid alterations in perilymphatic fluid pressure But for this to occur the cochlear aqueduct should be patent or there should be an underlying

congenital abnormality of middle or inner ear. (Parisel & Bukea) Goodhill & Simmons explanation for the occurrence of sudden deafness in above cases is due to rupture of labyrinthine window membrane due to pressure exerted on it. This rupture in turn leads to exertion of pressure on reissuers, basilar and tectonal membrane, which may rupture if pressure is great, which leads to mixture of peri-lymph and endolymph. So partly hearing loss is due to actual tissue injury at the site of break and partly due to mixture of the fluids.

Morrison ('75) views that there are no more facts to aid in deciding upon pathogenesis. Both sexes and all age groups were equally involved with no particular seasonal incidence.

Histopathology:

The pathology involved in sudden deafness has been reported by a large number of authors. The pathological changes occurring in the cochlear end organ secondary to sudden deafness, as reported by Schuknecht ('62) Beal('67), Joseph ('75), Tomoyukr ('78), Lindsay ('73), Nomura ('76),Isamo & Sando ('77), David ('67) are that the organ of

corti is often missing in the basal turus and individual hair cells tend to be missing at higher turus. Ganglian cell population is decreased at the basal tura but being normal towards the apex. Stria vascularis and toctonal membrane tended to be atrophied and rolled up, while being ensheathed in a syncytiumi of cells on the gaibus. Reissuers membrane would be collapsed and adherent to the basilar membrane. Saccule would be involved, while utricle and the semi circular canals escaped any damaged.

Schuknecht's ('73) report of pathological changes of 8 temporal bones in sudden deafness cases is given on the next page:

ORGAN OF CORTI

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

Hiroaki('78) performed electro cochleoGraphy on patients with sudden deafness. Based on the wave form pattern i.e., action potential and summatinG potentials he deduced the pallio-physioloGy of the disease. The wave form patterns were:

l.A.P. High response:- Where the A.P. response had normal or greater amptitude in 'H' part and absence in 'L' part of the wave.

2.Decrease A.P. high response:- A.P.amplitude in 'H' part was decreased and absent in 'L' part.

3.A.P. low response:- delayed latency in 'L' part even at maximum intensity.

4.- 3P + SP :- S.P. response had + ve ol - ve deflection.

6. AP & SP no response.

Impaired regions of sudden deafness were deduced by theabove 2 check patterns.

1.Temporary functional impairment of senaory cell and auditory nerve:-

i.e., patients Type 1 and 2 group showed improvement. Here, as the C.M. were recorded satisfactorily at time, it was unlikely that hair cells and auditory nerve would be impaired organically, but on the other hand it was a temporary change due to reversible physical or chemical change at the neural level.

2. Impairment at neural level:-

Here C.M. was normal and AP had no response. The C.M. response revealed that hair cell impairment was not serious but only impairment of the auditory nerve existed.

3.Organic impairment of sensory cells on auditory nerve:-

Type 3,4 and 5 groups showed slight or no improvement, so it was speculated that severe organic impairment of hair cells and auditory nerve existed.

The authors have also postulated that impairment at neural level suggested viral neuronitis or ganglionitis. While decreased AP high response, AP low response and AP-SP no response with depressed CM response suggested both viral endolymphatic labyrinthitis and neuroganglionitis;

Kiniura & 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 turus 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 generalised destruction of the inner ear structures and fibrous tissue invasion and finally complete ossification of the cochlea. Alford('65) & Supa('70) also came with similar reports.

Besides the earlier mentioned pathologic changes a leukemic patient has leukemic haemorhage

into bothperilymphatic and endolymphatic spaces in cochlear and vestibular systems. Endolymphatic hydrops were seen in cochlea andsacculus and vezti-bular aqueduct and endolymphatic sac was straightened. (Isamu '77).

CHAPTER V

CLINICAL FEATURES (SIGNS/SYMPTOMS).

The sudden onset of symptoms is the outstanding clinical feature of this type of deafness. Tinnitus is most frequently reported as being a presenting symptom (Grace ('76), Morrison('75) Snow('73) . It may be of varying degrees, which may either proceed the hearing loss (Morrison '75) or coma on simultaneously with the problem (Snow ('73) or may develop hours or days after the deafness has appeared. It is usually of a roaring quality, although noises like tickling bells have also been mentioned (Jaffe). It often subsides within one month, but may persist and even loverlast the deafness. Morrison '75 is of the view that presence or absence of tinnitus does not affect prognosis, although Frederick ('79) says its presence is indicative of poor propnosis. Mironeuko ('76), Schuknecht ('75), Goodhill ('72) have all reported tinnitus as being a very frequent symptom in sudden hearing loss.

There is considerable variation in the incidence of vertigo; and it forms an inconsistent symptom in sudden deafness (Saunders '71). In general about 40% of the patients have mild or transient vertipo and 10%? have incapacitating vertigo which lasts for 4 to 7 days, after which it may exist in milder degrees. It may accompany the hearing loss or follow it by hours or days or even years (Joseph '75). It can be the only symptoms in some cases (Goodhill '79), and may vary from a mild, giddy feeling to severe true vertigo with spontaneous mystagmus nausea and vomitting.

The frequency of vertigo seems to be related to the site of the pathology. In those with an end-organ localization or sub total deafness, acute vertigo is often a prominent symptom, it is less likely to be seen in truly sensorineural lesions and is more unusual in retrocochlear cases. To support the above statement, Morrison ('75) has given the following table.

Localisation	Males	Females	Mean age	Vertigo	Tinnitus.
Sensory (14 ears)	10	4	44	71%	86%
Total or Sub;total (9 ears)	5	4	45	63%	44%
Sensorineural (16 ears)	8		39	37%	44%
Neural 0(20 ears)	9	11	34	10%	90%

Vertigo does effect the prognosis inso far as cochlear lesions are less likely to respond well to treatment. Saunders('77) reported that if the deafness was associated with vestibular disturbances then, the symptom of vertigo with a downward sloping audiogram was a significant prognostic factor. Failure of hearing was strongly associated with severe vertigo. In cases with a flat or upward sloping audiogram, vertigo was a irrelevant for the prognosis.

Yoshida('7l), Nomer('72), Hozawa et al('76) and Emagi('76) have all reported ofoccurrence of vertigo with sudden deafness.

Deafness and audiologic studies:-

A great deal of variations occurs in the evolution of the symptoms of sudden deafness. The deafness 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. In most cases, onset can be dated exactly by the patient. He usually first notices either a stuffy sensation or tinnitus in the ear. Hearing loss may be noticed thep, being associated with a cracking or popping sensation. When annoying tinnitus occurs with deafness, the patient may even fear that he has had a stroke.

Hearing loss is often noticed on awakening in the morning when hearing has been perfectly normal the night before. Some patients are awakened from a sleep by the associated tinnitus. Less often, the hearing loss is noted during any of the days activities, such as those involving physical or emotional strain or even while at rest. Simmons ('77)[137] studied 89 patients with sudden deafness, some of the items from the

patients history were as follows:-

Present on awakening	- -	33
Exertional	-	24
Sedentary	-	15
dating	-	5
Not instant loss	-	5.

If the hearinp loss is bilateral, it will be noted promptly once interpersonal communication is hindered. On the other hand, unilateral losses may escape the patients detection until some specific test of the affected ear, such as use of telephone occurs. Dating of the onset is often difficult in children and in adults as well.

Usually the hearinp loss draws the patients attention promptly regardless of the presence or absence of tinnitus. The patient is often aware of the profound loss of discrimination that is initially presented. Unusual sensitivity to intense sound and displacusis are rare. As a rule, difficulty in localizing sound, is experienced.

Though different patterns of pure-tone audiogram are encountered, there is a general uniformity. Sudden sensory losses are mainly flat or show a high tone cut-off; sensori neural patterns are almost all high-tone; retrocochlear ones may be low-tone, trough-shaped, flat or high toned (Morrison '75). Initially recruitment is usually not present, but it may appear later if the loss of hearing becomes permanent(Snow & 73). In the permanent losses, recruitment is present in less than half the patients (Van Dishoeck'57[201] & Verger '61[200]).

The results of the investigations that are most frequently carried out with cases of sudden deafness are-audiometric loss is flat (moderate or profound), ABLE +ve, SISI +ve,Bekesy type 11 or Type III. Tore decay -negative, speech discrimination upto the SN loss. Vestibular functions are rarely involved, Shaia et.al ('76)[88]reported the findings in l220 cases of sudden hearing loss. The audiometric patterns as recorded by them are as follows:

I	Slope of audiogram	Incidence
	Low tone	12%
	Flat	32%
	High tone	31%
	Profound or total loss	25%
II	Bekesy patterns	Incidence
	Type I	22%
	Type II	59%
	Type III	11%
	Type IV	8%

Jerger ('60)[202] reported that the majority of patients with permanent hearing loss of sudden onset had type III Bekesy tracing. On subsequent study Jerger ('61)[200] reported that two thirds of patients with permanent hearing losses of sudden onset had patterns of response generally associated with a neural hearing loss. (low SISI scores, type III Bekesy audiograms, non-measurable discrimination scores and minimal recruitment) while one third had patterns associated with cochlear hearing loss (high SISI

scores, type II Bekesy audiograms, some recruitment and measurable but low discrimination scores) and accordingly they have been divided into cochlear and neural (Harbert'64).

Saunders('77)[138] has provided a table under asimilar division of sudden deafness patient.

	Cochlear	Neural
Bekesy	Type II or I	Type III or IV
Tone decay	Minimal	Marked
Discrimination	May be present	Absent
SISI	High scores	Low scores
Audiogram	Low tones partially presented	Flat of tough shaped
Recruitment	Present	Absent.

Based on the above factors Graham et all were able to localize the site of lesion as cochlear or retrocochlear in 30 patients. They also used electro cochleographic results to confirm their diagnosis in the above patients, Ecochg agreed with the diagnosis in 23 patients. Their diagnosis of the site of lesion as determined by Ecochg is as follows:-

Cause of deafness known	Total	Cochlear	Retro cochlear	Cochlear + Retro-cochlear	Normal
Neuroma	6		6		
Skull practice	4	3	1		
Mumps	4	2	2		
Chronic myeloid Leukema	2	1			
Typhoid	1		1		
Gentamicin overdose	1	1			
Labyrinthine membrane rupture	1	1			
Meniere	1	1			
No hearing loss	3				3
	23				

Stephens et al ('67)[94] reported a patient in whom a type IV Bekesy audiogram was found during the acute phase; withthe other tests indicative of neural lesion, but the type IV Bekesy audiogram then converted to Type II within 5 months with the other test results

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

Based on the audiologic test results, Beeningham ('62)[100] has differentiated between the sudden loss occurring 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 characterised by a flat curved, puretone audiogram with positive recruitment. The second one is a high frequency loss with no vestibular vertigo accompanying it.

Neill ('60)[162]reported of a typical pattern of audiometric test response, obtained with cases of sudden deafness associated with mumps. BC ths were characteristically lower than AC ths. speech reception thresholds were higher than the average puretone loss, discrimination scores were very low and there were indications of recruitment.

Kikawada('79)[163] has reported of a long term follow-up study of unilateral sudden deafness patients. It was performed in 28 cases who were diagnosed as having sudden deafness 10 years earlier. 18 cases showed no changes on audiograms, 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 3 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.

Vestibular signs are evident in the majority of patient with vertigo and in a few with no vertiginous symptoms. Approximately 50 percent of patients with 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)[179]

Vestibular symptoms accomtpanying sudden deafness as given by Shaia ('76)88.

Vestibular symptoms	Incidence
None	60%
Initially only	23%
Severe	10%
Persistent disturbances	18%

Spontaneous nystagmus, after abolishing fixation is usually seen to the opposite side, provided the patient is examined within a few weeks of the onset. Caloric responsesvary from normal to absent with all the usual modifications. Minor alterations in taste and lacrimation,on the affected side have also been encountered (Morrison '75). Cochleo vestibular symptoms and findings of 9 cases tested by Iain et al ('71)[127].

Cochleo Vestibular symptoms and findings in 9 cases.

Cases	Hearing loss Unilateral	Bilater	Hearing Improvement	Vertigo	Caloric test
1		Total	None	None	Not tested
2		40 to 50%	9 to 21%	"	"
3		45%	None	Yes	No response
4		68 to 70%	None	None	No Response
5		Total	None	None	Normal
3		62 to 69%	None	None	Normal
7		Total	None	Yes	No response
8	Total		Complete	None	Normal
9	Total		None	Yes	Ipsilated Canal paresis

Singleton et al ('77)[113] evaluated 51 patients suspected of having a perilymph fistula. They postulated that predominantly vestibular complaints had unrecognised perilymph fistula. Positional nystagmus was found to occur in supine, right ear 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) & (Hyder '79)[157] and in acoustic neuroma.

Depending upon the vestibular symptom Kirikae et al ('64)[25] divided their group of sudden deafness patients into two, the 1st first had high tone loss, with directional changing, positional nystagmus and the second group had hearing loss over the whole range with directional nystagmus towards one side. This difference was supposed to be due to quantitative factors of a lesion in the vestibule.

Pain is an occasional feature in suddenidiopathic deafness like, the pain sometimes 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 heading loss (Morrison '75)[136]. A sensation of pressure in the affected ear is experienced by many of these patients (Snow &'73)[179]. Headache is occasionally encountered and symptoms of viral upper respiratory tract infections occur as frequently as in 25 percent of the patients insome series. Fever, usually of mild degree may be present. Generally, however, 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 foundelevated white cell count in 59% and elevated erythrocyte sedimentation rate (ESR) in 41% of the cases with sudden deafness.

Age and sex distributions are fairly equal. Sudden sensory deafness is possibly commoner in males. The ages of these patients range from adolescences to 72 years (Morrison '75)[136] with 30 to 60 years being the most crucial range (Simmons '73). Nadol ('75)[76] reports that sudden deafness occurs most often between 8th to 20th years of age. Shaia ('76)[80] in his study of 1220 cases of sudden deafness condluded that there were slightly more females than males, although the difference was very small. Three-fourths of his patients were 40 years of age or older at the onset of the symptoms. He has also given a table.

	Incidence
Under 30 yrs	13%
30-39	13%
40-49	21%
50-59	22%
60-69	18%
70 Yrs +	13%

Morrison ('75) also reported that the retro-cochlear lesions were commoner in the younger age group. He also felt that in discussinr '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 actiology. Between '68 to '73, with a number of scattered exceptions, sudden deafness had started in every calendar month of every year and no more than 3 cases had commenced in the same month of the same year (Morrison '75).

Spontaneous Recovery:

Schecides ('73)[203] considers that in 40 -60 percent of patients with sudden deafness we can expect a spontaneous recovery. Singleton('71) stated that 25% recover completely, 25 percent show some improvement and 50 percent have a lasting profound hearing loss.

Moshe et al ('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., 8KHz it is 25%.

Spontaneous recovery is likely in patients with psychogenic deafness and it occurs very frequently in cases with multiple sclerosis. It is seen in a small percentage of patients with sudden loss due to hydrcps, whether the hydrops be idiopathic or due to syphilis or other diseases of the otic capsule. Reversibility is a feature of quinine and salicylate ototoxicity. There is also no doubt that degrees of spontaneous recovery are seen after definite viral infections such as zoster, mumps and measles, though it is unusual for this recovery to be complete. In patients who have vascular lesions minimal improvements inthe auditory thresholds can probably take place without specific treatment.

Secretory otitis media is a very common cause of sudden unilateral or bilateral deafness in all age

groups. Here there is little diagnostic difficulty, although some times the diagnosis can be missed without a typepanogram. This situation must account for the occasional "sudden sensory deafness" which recovers spontaneously. If the stapedial reflex is used for localisation purposes, this fallacy will be avoided. (Morrison'75)[136]

Morton et all ('66)[146]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.

Prognosis:-

The prognosis for the recovery of the hearing in sudden deafness is not so bad as is generally supposed. Approximately one third of patients have a return of normal hearing, one third are left with a 40 to 80 dB speech reception threshold and one-third have total loss of useful hearing, (Snow'73)[179]. Shimozaki ('74)[35] followed up cases of sudden deafness and analysed them for symptoms, sex, age, method of treatment, audiogram types and prognosis. He divided

the prognosis into (1) complete recovery (11.1%) (2) clear recovery (16.7%) (3) slight recovery (33.3%) and (d) unchanged (38.9%). The possibility of hearing improvement was found to be limited to one month from the onset of hearing loss. He therefore emphasized an "early treatment."

Morrison ('75)[136] is of the view that prognosis is poor insudden total deafness, not very favourable in sensory andsensorineural hearing loss, but excellent in neural Lesions. He provided a table for the cases which he tested.

Diagnosis	Treated	Responded	No response
Vascular lesions (15 ears)	10	4 (all partial)	6
idiopathic sensory (14 ears)	7	3	4
idiopathic sub-total or total(9 ears)	5	1	4
idiopathic sensori neural (16 ears)	6	3	3
Idiopathic neural (20 ears)	14	12	2

Snow ('73) postulates tha+ prognosis cannot be easkly related to the rapidity of onset, the presence of tinnitus - recruitment or type of Bekesy audiogram. The longer the delay between the onset of deafness and the onset of recovery, the worse the prognosis for complete recovery. But Simmons ('73)[130] 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. He has also given a relation between onset of symptoms and days of first audiogram in the form of a table:

Days of onset	Recovery good	Some recovery	No recovery
1	4	2	
2	5		1
3	4	1	2
4-5	1		
6-8	2	1	3
9-14	2	5	2
15-21		1	1
22-30			2
30 +			6

The audiogram curve is also found to carry some prognostic value, (Emmett et al '79)[103] 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 complete loss of 8KHZ. Speech discrimination valueis found to have no prognostic significance. Simmons ('77)[131] says that the threshold at 8KHz carries a prognostic significance. If the loss is either improving or stable at 8KHz, the prognosis is good or complete recovery is seen in 78% of cases. If there is no hearing at 8KHz, regardless of hearing at other frequencies, prognosis is 29%. A flat hearing loss has a better prognosis if the threshold is no worse at 8KHz than is at other frequencies. A worse prognosis is there is an additional loss at 8KHz.

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 '73). 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 vertigo. (Fortbild '65)[129]. Hypc-active caloric responses predict a very poor prognosis for higher frequency recovery and a normal caloric response may correspond with a better prognosis (Simmons '77)[137]. Nevertheless, complete recovery of hearing does occur at times even after several weeks of profound loss. Yoshida et al ('77)3 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, delayeddiagnosis. (Frederick '77)[144], Sinmons '77)[137] and Shaia et al ('76)[24].

Feature which aids inbetter prognosis, are time . between onset and diagnosis, hearing at 8 KHz, better speech discrimination scores, absence of vertigo and early treatment, (Simmons ('77)[137] , Shaia et al ('76).

features which make no difference in nature and outcome of sudden deafness are: a history of antecedent upper respiration infection; time of the year, treating physician and drug treatment given(Simmons '77)[137] .

Hiraki et al ('76)[137] performed electrocochleography on 34 patients with sudden deafness. The various wave form patterns which they got were (l) action potential high response (2) decreased action potential high response (3) action potential low response (4) Dominant negative summating potential (5) Positive or Negative summating potential (6) action potential and summating potential no response.

15 patients had complete recovery. The ecochg gram showed dominant - resummating potential wave forms in 8 of them, and action potential high response wave forms in7.

In 9 cases there was slight improvement. Here the findings were: Action potential low response in 2; positive summating potential in 4, negative summating potential in 1 ; action potential and summating potential no response in 2.

In 10 patients of no change: Action potential and summating potential no response was seen in 7 cases. Negative summating potential in 1 and decreased action potential high response in 2 cases.

So the authors concluded that Ecochggrams can be used to estimate the prognosis of sudden deafness in initial stapes. Similar results have also been reported by Nishida ('77)[71].

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.

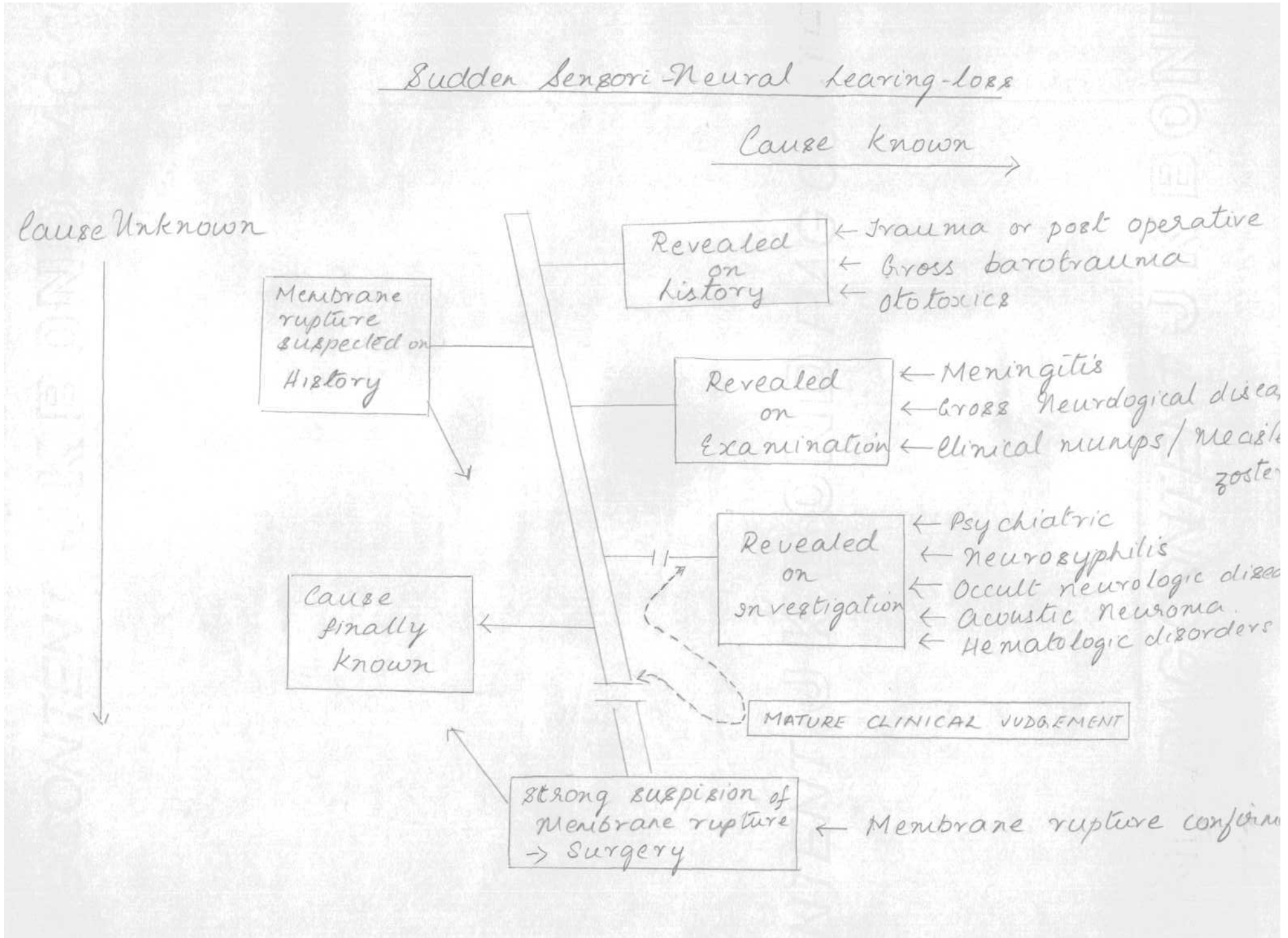
Anthony's('78) finding that, ina large treated aeries, the recovery rate was 87% for those seen at three days or less, 75% at up to one week, 53% at up to one month, 16% at up to three months, and 10%. if seen and treated at over three months from onset, Similar findinps have been reported by Shaia et al ('76). Rubin('68)[212] found that prognosis depended upon severity, none of hissevere cases showing any improvement.

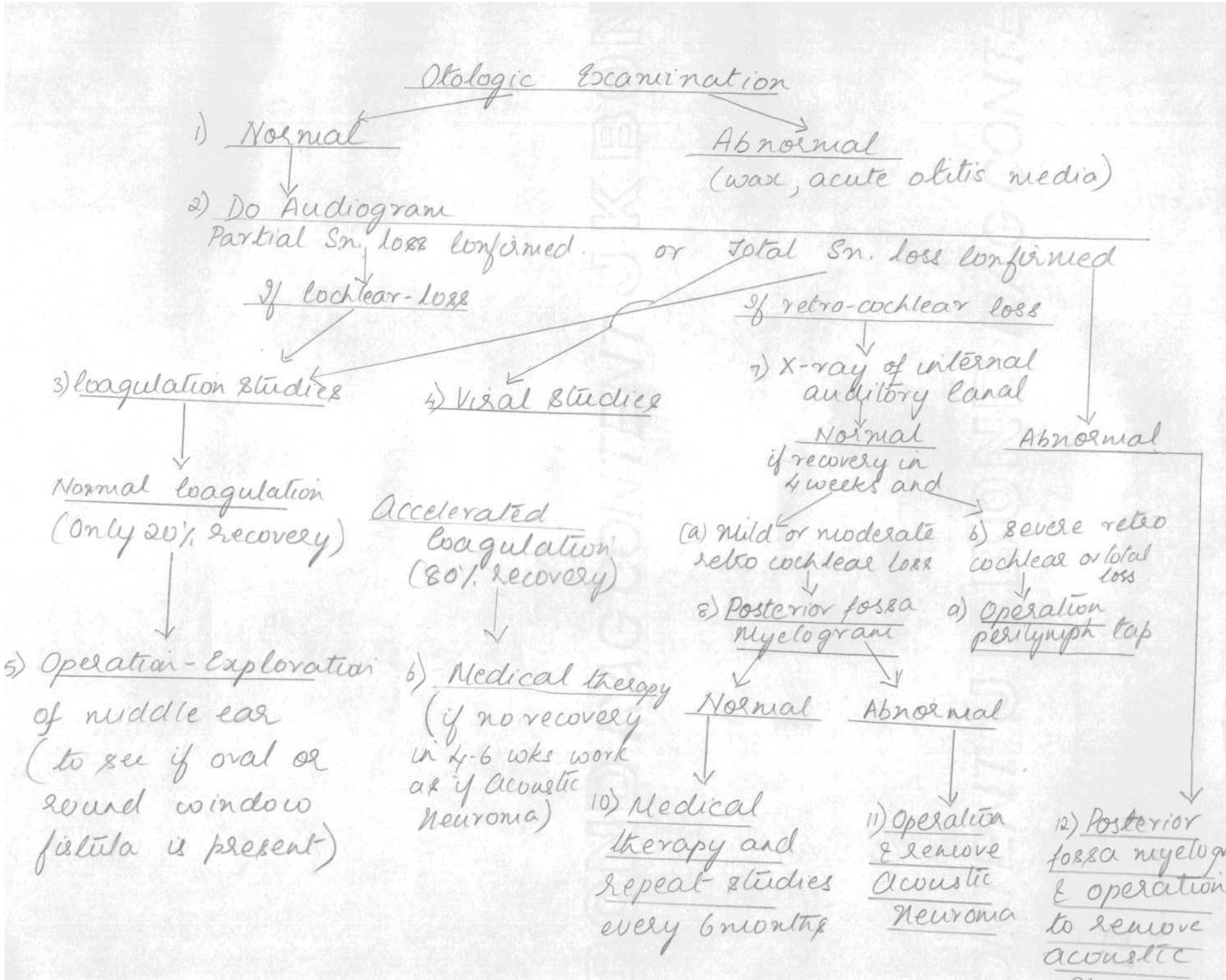
CHAPTER VI

DIAGNOSIS.

The problem of sudden deafness does not lend itself in any way to either a set routine pattern of special investigations for every patient, or to the "flow chart" approach in which, as in group analysis in inorganic chemistry, two alternative courses of action hinge upon the results of one test. There are a large number of diagnostic possibilities; some require invasive Techniques to establish, and others will be immediately apparent on talking to or examining the patients. Considerable clinical judgment therefore needs to be exercised in the investigation phase.

Richards('78) has given an illustration of the way in which some of the diapnostic groups are separated off early, and some late.


Sudden Sensori-Neural Hearing-loss
Cause known
Cause Unknown
Revealed on History
← Trauma or post operative
← Gross barotrauma
← ototoxics
Membrane rupture suspected on History
Revealed on Examination
← Meningitis
← Gross Neurological disea
← clinical mumps / Measle
zoste
Revealed on investigation
← Psychiatric
← Neurosyphilis
← Occult neurologic disea
← Acoustic neuroma.
← Hematologic disorders
Cause finally known
MATURE CLINICAL JUDGEMENT
Strong suspision of Membrane rupture -> Surgery
← Membrane rupture confirm


Otologic Examination
1) Normal
Abnormal
(wax, acute otitis media)
2) Do Audiogram
Partial Sn. loss confirmed.
or
Total Sn. loss confirmed
If cochlear-loss
If retro-cochlear loss
3) Coagulation studies
4) Viral studies
7) X-ray of internal auditory canal
Normal
if recovery in 4 weeks and
Abnormal
Normal coagulation
(Only 20% recovery)
Accelerated coagulation
(80% recovery)
(a) Mild or moderate retro cochlear loss
b) Severe retro cochlear or total loss
8) Posterior fossa Myelogram
9) Operation perilymph tap
5) Operation - Exploration of middle ear (to see if oval or round window fistula is present)
6) Medical therapy
(if no recovery in 4-6 wks work as if Acoustic Neuroma)
Normal
Abnormal
10) Medical therapy and repeat studies every 6 months
11) Operation & remove Acoustic Neuroma
12) Posterior fossa myelogr & operation to remove acoustic

Althouph a basic identification of sudden deafness is most frequently made without much of a - problem, a definite diagnosis of the condition lies a long way off, for which, a detailed otolopic and general medical histories and physical examinations are necessary. The above given steps put forward by Jaffe should be proceded in an orderly fashion in order to arrive at a correct diagnosis, while doing so, one should constantly keep in mind the different possibilities that could eventually lead to sudden deafness.

1.Otology:

A detailed otologic history should be obtained concerning previous ear problems or surgery, previous ear examinations, vertigo and/or tinnitus. Enquiry is also made into the possibility of trauma or noise exposure. Whether a family history of deafness exists is also important to enquire.

Besides otologic history a general medical history is also very important. It should include previous general medical problems, cardio-circulatory disease, anti-coagulant drug intake, ototoxic drung

use, recent viral or bacterial upper respiratory infection, related infections or contacts and recent unexplained fever, chills or malaise.

Otolopic examination:

Sudden deafness may be due to sudden onset of a conductive or sensorineural hearing loss. A conductive hearing loss is often accompanied by some obvious problems likewax in the eternal auditory canal or an abnormal tympanic membrane as in serous otitis media which can be identified by otoscopy. Eustachian tube obstruction is commonly identified as a cause of sudden deafness. Unless signs of eustachian tube obstruction are obvious, prominent short process of malleus, fore shortened and horizontally positioned handle of malleus, retracted tympanic membrane or poor mobility or penumatic otoscopy should be identified, Eustachian tube obstruction secondary to the upper respiratory infection should not be indicated as a cause of sudden deafness. When tympanic membrane is normal, a Sr, loss is most likely present, then you should proceed to testinp hearing.

The medical examination should include a search for bradycardia, airhythania and other cardiac

and circulatory problems. Buils,urine and blood studies should be done which should also include sedimentation rate, blood count FTA and ABS tests, special studies for hyper co-agulation, lipidemia syphilis, hyper-viscocity and macro glabulinemia and other syndromes should be included. This is done to elicit possible clues to the etiology. Charachon et al ('74)[80] have stressed the importance of electrophoresis in the dia-gnosis, as this would help in discovering myeloma, if present, which is an indication of poor prognosis. After this proceed to audiologic testing.

2. Audiogram:

Initial audiogram gives idea ofquantity of loss, also helps to know the improvement and in determining prognosis; Pure tone audiogram along with speech test-ing will help to differentiate between cochlear and retrocochlear for a more definitive and an accurate differential diagnosis Siegel et al have recommended a battery of tests, which although are time consuming can be selected depending on the individuals needs and necessities. These are:

1. Tuning fork tests - Rinne & Weber.
2. Whispered voice test.
3. Pure tone air and bone conduction.

4.Speech reception threshold.
5.Speech discrimination threshold.
6. Modified tone decay test.
7.Loudness recruitment (SISI & ABLB)
8.Bekesy audiogram.
9.Binaural pitch matching.
lO.Impedence measurement.
11.Other tests such as DL for frequency, temporal integration; competing messages, Stenger.

Snow ('73)[179] recommends the determination - of speech reception and discrimination scores initially and at two to three days interval until the hearing loss stabilizes. He also stresses that, of particular interest throughout the illness are the tests for recruitment and tone decay, the SISI and Bekesy.

Along with audiologic evaluation, the vestibular evaluation should also be done. In general vestibular studies with the use of the Fitzgerald-Hallpiketest with electronystapmographic monitoring are of interest but need not be carried out in the acute phase.

Siegel has recommended the following vestibular tests to be done with sudden deafness patients: (1) Rhomberg, tandum standing test (2) Gait testing (3) Spontaneous nystagmus tests (4) Positional test (5) Caloric test (6) Electromyographic test.

According to the earlier given protocol (by Jaffe), if cochlear loss then proceed to No.3 & 4. If retro-cochlear proceed to No. 7. If loss is total this differentiation is not possible which would otherwise be possible at this level. For such conditions evaluate as if they were present in both the categories, i.e.No.3,4 and 7.

3.Co-agulation Studies:

The sudden deafness may suggest a sudden vascular occlusion which could arise from thrombosis of cochlear vessels. There are many patients with the above evidence. Therefore, the possibility of a thrombosis arising from a hypercoagulation state was considered and multiple tests have since then been utilised to assess the

presence of accelerated coagulation . When coagulation is normal - proceed to No.5. When coagulation is accelerated - proceed to No.6.

4. Viral Studies:

About 1 out of 3 patients with sudden deafness will have an antecedent upper respiratory infection which can be identified by the above investigation. In addition to these overt infection the viral studies will identify other patients with sudden deafness who have had a recent but subclinical infection. Luchmann('74)[81] has especially stressed on viral investigation to determine whether post-influenzal disturbances are eucephilitic or otitic, here he says that vestibulometry and other tests should be used.

About 60% of the patients with viral infections will recover spontaneously and no specific antiocial therapy exists for them. Medical therapy is indicated so proceed to No.6.

5. Operation:

Operation is done to explore the middle ear and to see if oval or round window fistula is present

as a definite pre-operative diagnosis of the fistula is difficult or impossible. Most authors therefore recommend surgical exploration of the ear if fistula is suspected. (Edmund &73, Farmer & Thomas &'76, Fee '68, Freeman '74, Goodhill '71, Healy et al '74, Mc Cormick et al '76, Mulany et al '74, Pullen '72, Stroud & Calcaterra '70). Spontaneous recovery occurs in these cases if complete bed rest is given. If vertigo subsides and hearing is restored,the surgical intervention should be postponed while he is advised to avoid valsalva manoevre, vigorous nose blowing etc (Gundersen et al '78)[154]. When vertigo does not subside, a surgical intervention is done wherein peri-chondrium autografts are placed on the fistulous area. (Goodhill et al '72)[139].

6. <u>Medical Therapy</u>: There are many types:

(1) Ambulatory care: where as vasodilating drugs such as nicotinic acid and roniacol have been used.

(2) Hospitalization: bed rest with intravenous therapy with histamine and low molecular weight dextran have been used (This topic will be discussed in detail under the chapter of Therapy).

7. X-ray views of internal auditorycanal:

A retrocochlear hearing loss may be due to compression of 8th nerve as in acoustic neuroma in internal auditory canal. As it grows, it produces erosion of the surrounding tone and this may be apparent on X-ray by comparing normal and abnormal sides. If the height of the internal auditory canal is equal on both sides, either no tumor or a very tiny one may exist. Petrous pyramid X-ray also helps to identify tumor. Its importance has been stressed by (Shaia et al ('76)[85], Dujour ('75)[39] Wilson ('73)[86] has also put forward the use of internal auditory canal roentgenograms and cerebello pontive-asterm myelogram for the identification of acoustic tumor. If X-ray is normal wait for 4 weeks. If no recovery and if hearing loss is mild-proceed to No.8. If hearing loss is severe or total proceed to No.9. If X-ray is abnormal then proceed to No.12. Jaffe has stressed that under no circumstances should the work up for a retrocochlear hearing loss stop with just X-ray of the internal auditory canal.

8. Posterior fossa-myelogram: It is used for detecting small acoustic neuromas. Contrast radiographic studies of the internal auditory canal may reveal a filling defect consistent with acoustic neuorama. Radiographic examination of the temporal bone and cervical spine are also indicated to avoid overlooking mastoiditis, primary cholesteatonia, acoustic neuroma, and spondylosis deformans. (Snow '73)[179].

9. Operation is done for perilymph tap.

l0.Medical therapy.

11 & 12. Operation to remove acoustic neuroma.

The above given protocol by Jaffe has covered up a large area of diagnosis, although a few other points have been put forward by others which should also be taken care of in the diagnosis of sudden deafness. One such diagnostic point is the lumber puncture,

Lumbar puncture:

Judgment must be exercised in the individual patient with sudden deafness in determining whether a lumbar picture 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 cerebellopontive angle tumors, meningoencephalitis and trauma (Snow '73)[179].

The other authors who have experimentally proved and pointed on the use of the above testing are Sawada, Kumangami, Jinnouchi and Mirouchi('76)[38].

Including all the above points, the National Registry for idiopathic sudden deafness at the Department of Otolaryngology has set up its diagnostic protocol (done by Siegel). It is as follows:-

I. History:

1. Otolaryngologic and complete general.
2. Have the patient fill out the sudden deafness questionnaire which is forwarded with other findings.

II. Physical Examination; Otolaryngologic and complete general.

III. Include consultation for:

1. Fundoscopic examination.
2. Neurologic examination -(Especially of the cranial nerves)
3. Medical evaluation for vascular, collagen or other systemic diseases.

IV. Audiologic evaluation:

1. Tuning fork tests: Rinne & Webe.
2. Whispered voice test.
3. Puretone air and bone condution.
4. Speech reception testing.
5. Speech discrimination testing.
6. Modified tone decay test.
7. Loudness recruitment test (SISI & ABLB)
8. Impedence measurements
9. Bekesy testing
10. Binaural pitch matching.
11. Other tests such as DL for frequency temporal integration, competing message, stenger.

V. Vestibular evaluation:-

1. Rhomberg, tandum standing.
2. Gait.

3.Spontaneous nystagmus.

4.Positional test.

5.Caloric test.

6.Electronystagmography.

VI. X-ray:

l.Skull

2.Mastoid and internal auditory meatus.

3.Chest.

VII.Cardiac testing:

Vital signs (Blood pressure, pulse, respiration, temperature).

VIII.Hematologic testing:

l.WBC count.

2.Hemoglobin count

3.bedimentation rate.

4.Platelet count.

IX. Co-agulation studies:-

1.Prothrombia consumption.

2.Prothrombia time

3.Partial thromboplastin time (PTT)

4.Platelet count.

X. Renal:

1. Urine analysis.
2. Creatinine.

XI. Endocrine:

1. F B S
2. P B I

XII. Biochemical:

1. Total protein
2. Albumin
3. Globulin
4. V D R L
5. F T A

XIII. Lumbar Puncutre:

1. Opening pressue
2. Colour.
3. Cell count and Differential
4. Protein.
5. Serology
6. Glucose
7. Viral culture.

XIV. Viral studies:

1. Whole clotted blood
2. Stools
3. C S F

After having done the evaluations given above it would be possible to arrive at a definitive diagnosis in most conditions. There have also been certain points put forward by different authors which along with the other evaluations, would make the diagnosis more valid and reliable. Some of the points are as follows:-

In idiopathic viral cases, the patient will most frequently come up with a history of 'flu', 'cold', 'upper respiratory infection, sore throat, sinusitis, allergy flare-up, virus exposure to family or work-related contacts with individuals who have had viral infection. Evenif the exposure antecedates the hearing loss by several weeks, the suspicion should never be overlooked.(Goodhill '80).

In idiopathic vascular cases the patient often gives a history of previous cardiac or hypertensive disease with or without anticoagulation therapy. There may be history of diabetes, arteriosclerosis, hyper-cholestemia, hyperlipdemia or other systmic diseases involving micro-vasculature.(Goodhill '80). A positive serologic test also suggests a vascular involvement (Gopichand '68)[132].

Jaffee '76 said that milder degrees of sensory neural loss, centering around 4 KHz is secondary to the transmission of excessive vibratory energy, akin to a noise-induced loss, whereas a diffuse pattern of loss seen with massive force are related to membranours tare and contusions.

In cases of sudden deafness due to head injury a haemorrhage behind the tympanic membrane will serve as a bellwether, always suggesting a temporal bone fracture. CSF otorrhea is common with transverse fracture. Here there is also an involvement of facial nerve. When a high frequency notched hearing loss exists then rupture of reticular lamina and detachment of the organ of corte from the basilar membrane may exist and the hearing loss is more or less localized at high frequencies. If injury involves the rupture of reissner's membrane, the oval window and/or the round window, the hearing loss is at low frequencies.

Simmons '68[148], Pullen '79[102] Goodhill '80 have arrived at the following points which suggest a membrane rupture. They are:-

(1) If the patient is less than 45 years of age, has never had attacks of dizziness or hearing trouble before.

(2) if he noticed hearing loss after hearing a loud 'pop' or a marked roared tinnitus.

(3) suggestive points are if on that day the patients' physical activity could have caused an increase in intrathoracic or intracranial venous or C S F pressure.

(4) if more than a modest amount of alcohol was drunk beforehand.

(5) when there is a history of antecedent sudden physical exertion,barotrauma or head injury,diving or, flying.

(6) recurrent bouts of labyrinthitis, meningitis or both are reported.

(7) when there are known temporal bones, cranial or extracranial skeletal anamolies.

(8) objective evidence of a persistent vestibular disturbance exists when other etiologic factors have been eliminated.

(9) audiometric curve has a prognosticative value.

(l) A history of trauma with sudden onset of dizziness and/or hearing loss.

In some cases the stress or exertion related to the barotrauma may be very irrelevant to the consequence, like hearing loss/allowing lifting coughing, sneezing etc. Since such conditions are not unusual, they may be completely ignored. Therefore a careful history and detailed questioning are required to elicit such possible cause-effect relationship.

Here audiogram will reveal S hearing loss of cochlear origin. If audiogram is normal, but you suspect a fistula than an audiogram with 10KHz increments testing from 400 to 1300Hz isdone, a hole in the audiogram will be present due to rupture of the intra cochlear membrane. (Kenneth et all '78)[114] .

Vestibular tests in These patients will reveal positional nystagmus with the involved ear down and a reduced vestibular response to calorie testing. But it is inadvisable to do intensive vestibular testing during the acute periods of sudden hearing loss as it causes stress on the involved ear (Sean '77)[89] .

Stroud et al ('70)[75] have given the characteristics of sudden deafness due to membrane rupture which are different from meniere's disease. In sudden deafness the age of onset is often less than 23 years; there is an abrupt onset occurring during a moment of increased intracranial venous pressure. Basically a fluctuating neurosensory loss of cochlear type with hearing loss more marked in the higher frequency. Discrimination scores are more depressed than the pure tone and speech reception valves. The results of SISI, ABLB and Bekesy testings are more typical of cochlear loss.

Differential Diagnosis:-

The differential diagnosis to be considered is implied by the clinical work-up. However, consideration should be given to certain disease processes for which definitive diagnostic studies are not available. Among these diseases are labyrinthine otosclerosis, bacterial labyrinthitis, cerebellopontive angle tumors, trauma, otoloxic drugs and idiopathic endolymphatic hydrops(meniere's disease) which can usually be differentiated, but on rare occasions may masquerade as sudden deafness.(Snow'73)[179].

Of these conditions it is the meniere's disease which is most frequently confused - on which a large part of the following discussion is concentrated.

Sudden unilateral sensory deafness and tinnitus herald the onset of idiopathic endolymphatic hydrops is about 3 per cent of patients with this disease(Morrison '75)[136]. Hallberp ('68)[148] reported that sudden deafness was the initial symptom in 4.4% of patients diagnosed as having meniere's disease.

The diagnosis of idiopathic endolymphatic hydrops is arrived at by exclusion of the known causes of hydrops and by careful consideration and elimination of the other likely diseases known to produce vertigo. The history is all important. Most patients with hydrops have a fairly classical story of intermittent tinnitus, fluctuating hearing loss, displacusis or episodes of distorted sound, dull mastoid or occipetal pain and paroxysmal attacks of vertigo lasting from minutes to hours if the attacks are severe enough. (Morrison '75)[136]. He has also given behaviour and progression of the hearing loss in menieres' disease patients(471 ears).

	No. of ears.	Percentage of total.
Fluctuating	240	51%
Progressive with fluctuation	120	25.5%
Sudden with fluctuation	48	10%
Progressive	39	8.5%
Sudden	24	5%

There are many signs and symptoms which are common between the two groups i.e, sudden deafness andmeniere's disease, which makes the differential diagnosis difficult. Lehndardt et al ('76)[72] have tried to answer the question of whether there is any relationship between the two conditions. They have argued as follows:

In cases of sudden deafness, some hearing generally remains. It can be the first symptom of meniere's disease, like the first attack of vertigo which may occur without deafness. Measurement of stapedius threshold and Bekesy audiogram, especially the difference between the impulse tone and the continuous tone, indicates inner hair cell damage and are helpful in diagnosis of sudden deafness.

But pathological adaptation is seenin both cases. A decrease in distance between the hearing threshold and thereflex threshold is a further indication of inner ear damage, which is also seen in both conditions. The independence of the stapedius threshold up to a hearing loss of 60 dB seems to be an indication for the differential function of both types of hair cells. The reflex is supposed to be dependent upon the presence of inner hair cells because the reflex threshold increases as soon as an inner hair cell damage of about 60 dB occurs. Thus, the authors concluded that the clinical and audiological findings reveal no difference between sudden deafness and menieres disease. So it is frequently the results of the neurological and vestibular examination which are made use of to differentiate the two.

Neurological signs and symptoms were used by Ishikowa et al ('76)[6] to differentiate the two conditions. Their results were: (1) meniere's disease was characterized by spontaneous (93%), paraxysmal 95% and recurrent(100%), vertiginous attacks accompanied by cochlear signs such as tinnitus (93%), hearing loss (73%), while in patients with sudden deafness, the

nature of the vertigo was provoked (40%) as well as spontaneous (32%), paroxysmal (88%), single attacks, vertigo was much commonly observed.

(2) In patients with meniere's disease, hearing impairment was ingeneral slight and was characterized by deficiency in detecting low pitched and middle pitched tones. Sudden deafness was characterised by highly impaired hearing i.e., total deafness was observed in 45%.

(3) Fluctuations of auditory threshold was observed in 29 cases (71%) regardless of the course of meniere's disease and these fluctuations were much more common in the low frequency range. In sudden deafness on the other hand, hearing impairment tended to improve in all frequency ranges.

Yamazaki et al ('76)[7] tried to differentiate the two conditions using the mild caloric eye tracking pattern test (MCETP-test) and RE-MCETP test, Such mild caloric nystagmus in patients of sudden deafness and meiniere's disease evoked super imposed ataxic eye-movements onthe eye tracking pattern as well as caloric nystagmus. These ataxic eye movements in the MCETP tests were apparently evoked with sympathetic enhancement of the labyrinthine in the autonomic

nervous system in the meniere's disease, unilateral cephalo-cervical autonomic nervous system in sudden hearing loss. This helped to differentiate between the two.

From the above results it would be possible to differentiate between meniere's disease and sudden deafness.

Fluctuant Loss:-

Although fluctuant hearing loss is more commonly encountered by the otologist in relation to middle ear disorders, it is well established that fluctuations in senaori-neural losses do occur. In menieres syndrome, from whatever cause, vertigo predominates over the other symptoms in the tetrad and the hearing loss simply means that this episodic disorder of the labyrinthine function has produced endolymphatic hydrops, with or without associated degeneration of the neuro-epithelium.(Hinchcliffe'73)[208]

Shea, however, uses the term 'fluctuant hearing loss' to describe an entity which is distinct from menieres' disease and consists of roaring tinnitus, fullness and fluctuation in hearing with or without vertigo; he finds it many times more common than the full tetrad of meniere's disease. Although

the hearing may return to normal after the first attack, fullness and roaring remaining, and becoming worse woth each subsequent attack 'Shea '75). He feels that multiple metabolic disorders underlie the condition possibly with a defect in endolymphatic drainage, and uses a complex schedule of investigation, and-in-patient treatment by bed-rest, non-smoking, low salt diet, oxygen inhalations and drug therapy.

Although fluctuant hearing loss may occur as a result of recognised clinical entities - in perilymph fistula after stapedecomy (Goodhill '67)[210], multiple sclerosis (Clemis '75)[211] and removal of a cerebral hemisphere glioblastonia (Hansen'73), the existence of an idiopathic entity, disti-nct from meniere's disease or syndrome and sudden deafness, need further clarifications.

CHAPTER VII

TREATMENT

The frequent spontaneous recovery of hearing to normal or near normal levels makes evaluation of any form of therapy of sudden deafness very difficult.(Snow '73)[179]. No controlled studies have been performed with any of the forms of therapy that have been advocated. 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 judge whether any form of therapy advocated for sudden deafness produces a higher recovery rate than would have occurred spontaneously.

Sudden deafness requires immediate investigation and treatment if there is to be any prospect of salvaging the hearing. It presents an otologic emergency (Jaffe) and a diagnostic challenge (Morrison '75)[136]. There is much to be said for admitting to hospital the patient who is seen within

three weeks of onset. After this time there is little likelihood of recovery and investigation can proceed at a more leisure pace. The sooner the treatment is given, the better the results will be; when the delay does not exceed a few days, good results may still be expected. After a week, it is still possible to obtain pood results, but this is no longer certain. (Guilton'65)[18].

One point that is interesting about these patients is that cases of sudden deafness apparently have very little tendency to relapse, at any rate, not within a short time. still a few cases of relapse have been reported, one of them after an interval of 15 years. By dietary measures and treatment to improve the balance of the neurovegelative system,anti-allergic treatment or endocrine treatment, the physician can try to improve the deficient properties of these constitutionally pre-disposed patients(Guilton '65) .

It must be emphasized that the condition of sudden deafness cannot be labelled idiopathic until recognizable pathologies have been excluded. If this is not done, a conflict between therapeutic and diagnostic process frequently exists and under such

circumstances Morrison('75)[136] prefers To err on the side of therapeutics. Therapy of course can be withdrawn in a few days once the diagnosis is established, whereas a few days delay may affect the ultimate result in sudden hearing loss. This is one of the occasions where slavery to diagnosis must be avoided.

If this policy is adopted it is essential to initiate some of the basic investigations prior to steroid therapy since the latter may affect the test results. This applies particularly to the taking of blood samples for a large variety of tests (e.g. for syphilis, viral antibodystudies, blood sugar analysis, renal or liver function tests, blood counts and E S R). If hydrops is suspected, glycerol dehydration and plasma osmolarity studies need delay therapy by only 12 hours, specialised tests for localization should precede treatment since incochlear lesions, there may be an advantage in adding vasodilator therapy. Other investigations such as vestibular analysis, polytomography and Ecog can follow the onset of treatment. Contrast, radiography, subsequently considered necessary in many of these patients, demands no urgency.

The therapies currently advocated includes vasodilation, steroids, anticogulation, reduction of the viscocity of the blood, sedation and trancuilization, vitamins, corticosteroids and bed rest etc. But one of the main factors that decides the therapeutic approach is whetherthe cause of the problem is 'known' or 'unknown'. In the group where the cause has been established (syphilitic tuberculous and acute inflammatory disease of the nervous system, neoplasms including acoustic neuroma, meniere's disease etc), the treatment problems are those of the individual therapy for the condition. The problem of the diagnosis of the membranerupture has beenhighlighted already and the final diagnosis can be made only by tympanotomy. The technique of repair has been described by Goodhill '76. In proven viral infections, Morrison et al('70)136 find it right to advocate the use of steroids but it has not been found to be of much help in cases of deafness due to measles or mumps which is of long standing nature. Unproven viral infections should be considered with the cause unknown group(3ichards'78).

Under the cause unknown group, the data are confusing and the treatment methods diverse, the reasons being: (l) that where etiology is in doubt,

treatment is empirical. (2) that several different pathologies may be grouped under this heading, each with a different prognosis and treatment response (3) that there is controversy about the natural history (4) that the effectiveness of some treatment methods has been doubted.(Richards '78).

Treatment Methods:

Vasodilation has been advocated byVan Dishoeck & Bierman ('57)201, Sheehy ('60)190, Jaffe ('67) Rubin ('68)[206] and Sheehy et al ('76)[80]. Van Dishoeck & Bierman mention spasmolytica but do not specify the agents, Sheehy advocates intravenous histamine phosphate initially, in 250 cc. saline at the rate of 50-60 drops per minute to be followed by intra-muscular histamine phosphate. He also advocated the use of intravenous procaine in cases who are hyper-reactors to Kstamine. Subsequently, oral therapy consists of sublingual histamine phosphate and nicotinic acid. Sheehy and Jaffe are also of the same view.

Rubin advocated hyoscine or atropine, intramuscularly or intravenously in the acute phase and

procaine hydro-chloride intravenously after several days or weeks. He also recommends nylidrin, a non-cetacholamine and B-receptor stimulant. He reported, return to normal hearing in 50% of his patients.

The results of vasodilation therapy appear to approximate spontaneous recovery rate (Snow '73)[179]. Study of the effect on cochlear blood flow in guinea pigs by vasodilating drugs and some related agents, was done by Suga & Snow, who showed that nicotinic acid, even in massive doses, has no measurable effect on cochlear blood flow. Histamine phosphate and, betahistine increased cochlear blood flow in dosages that produoe bronchospasm in the guinea pigs and may well produce vasodilation on the basis of anoxia.

The rationale of vasodilation therapy for sudden deafness is questionable in view of the pre-ponderance of evidence for the viral etiology of most cases of sudden deafness. Some now advocate vaso-di-lation 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 micropetechiac in viral infections. Resolution of the question of the efficacy of this therapy must await well controlled clinical studies. (Snow '73)[179].

Morrison & Booth ('70)[193] 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 prednisone was reduced to 25 mg. and thereafter to 20 me. daily. Steroids were 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. The authors have not reported of any cases of relapse. Pregnancy diabetes and hypertension have been contraindicated. Morrison & Booth ('70)[193] have also tabled the responses of the cases to treatment.

Diagnosis	Treated	Responded	No response	Treatment	Not treated
1. Vascular Lesion (15 ears)	10	4 (all partial)	6	Various vasodilators	5
2. Idiopathic sensory(14 ears)	7	3	4	Steroids with or without vasodilators	7
3. Idiopathic subtotal or total (9 ears)	5	1	4	Steroids with or without vasodilators	4
4. Idiopathic sensori neural (16 ears)	6	3	3	Steroids	10
5. Idiopathic neural(20 ears)	14	12	2	Steroids	6

Some of the newer corteo steroids which have been used in the treatment of sudden deafness are dexamethasone, paramethasone, triamcindone, 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. The earlier the treatment is begun the better the results. Morrison & Booth ('70)[193] have also illustrated the effect of time delay in steroid treatment in the below table:

Localization	Treatment started within			
	1 week	1-2 weeks	2-4 weeks	1 -3 months
Sensory(7 ears)	1 good recovery	2 partial recovery	2 no recovery	2 DO recovery
Total or Sub-total(5 ears)	1 partial recovery 1 No recovery	1 No recovery		2 No recovery
Sensorineural (6 ears)	1 Good recovery	1 partial recovery 1 no recovery	1 partial and 1 No reco-very	1 No recovery
Neural (14 ears)	6 Excellent recoveries	4 excellent recovery 2 partial	1 No recovery	1 No recovery
Totals	90% improved	82% improved	20% improved	0% improved

Steroids with anticoagulants have also been used like the adreno cortico tropic hormone (ACTS). Hartley & Coleman ('74)[147] used this on a few cases along with nicotinic acid, prednisone and chloretrimeton. It was used for 10 days and pre-treatment hearing level was observed. So they have attached an importance to ACTH. Saunders has noted the use of ACTH in the viral and vascular etiology of sudden hearing loss, wherein viruses attach themselves to red blood corpuscles and produce hemagglutination and has reported improvement. Bolognesi ('60) used ACTH with a case of hypercoapulation and found complete recovery. Schiffet al('74)[147] has listed the several effects ACTH can have, some of which explain the prompt recovery.

1. AGTH: is a stimulant for corticosteroids from the adrenals.

2. It tends to suppress antigen -antibody reaction although it does not inhibit its response. The union of antigen-antibody is not prevented, nor is the release of histamine from sensitized cells prevented. Little protection is offered against anaphylaxis however, the intensive response is suppressed and its major therapeutic benefit is

atributed to suppression of inflammatory response after cellular injury.

3.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.

4. The mechanism of action of ACTH on its receptor cell is to stimulate the formation of adenyl cyclase which is a messenger from the endocrine gland to the cell interior. This in turn releases the amount of cyclic AMP from the ATP that is found within the cells.

5. 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 rouloux formation with occlusive effect on the small vessels in the periphery. ADP has a characteristic of increasing, the stickiness of platelets.

Cyclic AMP is increased by the beta ceta cholomines and is decreased by alpha catecholamines.

Selective action on beta sites produces vasodilatation and bronchodilatation by virtue of its action on smooth muscles. Anything that antagonizes alpha-catecholomines would be for the betterment of an embarrassed circulatory state (Schiff et all '74)[147] . Ishigama et al (& '76), Jakobi et al ('75), Meyerhoff ('79) have all given evidence to support the recommendation of ATP in the treatment of sudden deafness.

The other drug to gain importance in the treatment of sudden deafness is Heparin. Bolognesi('60)[207] advocated anticoagulant therapy after improvement in the hearing was seen in 3 of 5 patients on heparin and coumadin therapy. Schiff et al ('74)[147] has listed the effects of heparin as being:

(1) it exerts its anticoagulant effect specifically as an antiprothrombin action which inhibits th- conversion of prothrombin to thrombin. It does not thin the bloodnor does it alter the sedimentation rate.

(2) It specifically stimulates lip protein lipase formation. This enzyme 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.

(3) The inhibitory effect on antigen-antibody reaction was noted by Drapstedt . This mechanism is related to the fact that heparin complexes and binds histamine 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 cell. This binding of histamine and heparin limits the cyto-destructive effect to the benefit of the capillary with its endothelial cells with other responsive cells.

(4) The anti-inflammatory effect, indicates that this highly sulfated acid mucopolysaccharides of the ground substances and connective tissue acts as a caution exchanger and complexes the basic amines and polypeptides.

Donaldson ('79)[124] used heparin with 23 cases of sudden deafness and found that 14 had complete recovery (60.4%), 2 had good recovery (8.7%), 7 had poor recovery (30.4%). None of the patients failed to recover.

Jaffe('67) advocates low molecular weight dextran to reduce the viscocity of the blood. It is administered by the intravenous route in 500cc. 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. (Meyerhoff '78) & '79)[111] & 110. as in other conditions and early diagnosis and treatment are considered to be crucial for successful therapy here also. The mean hearing gain in those patients who came for dextran treatment later than 1 week, was less than half of the gain that resulted once treatment had begun within the first week. (Otto et al '76)[54]. Spoendlin & Rossberg ('77)2 are of the view that a more significant improvement is obtained when paparin is also administered along with dextran.

Shea et al ('77)[131] evaluated diatrizoate meglumine (hypaque) in the treatment of sudden deafness. They treated 30 patients with hypaque and a vasodilator regimen. Of these patients 30% had a good response, 23% had a moderate response and 47% had no response.

The same authors did another study in '79[103] wherein they treated 31 patients of which 12 had complete recovery. Shea ('77)[131] has quoted Prof.Morinitsu who reported that 22 of the 60 patients with sudden deafness were treated with hypaque and had complete recovery of hearing. The explanation put forward by him is that sudden deafness is due to a breakdown in the blood cochlear barrier in the area of the stria vascularis with the subsequent decrease in the endocochlear DO potential. Because of the molecular size and configuration of the diatrizoate meglumine, the broken membrane pores are filled and the sodium pump is activated to restore the normal DC potentials.

Stellate ganglion is made up of inferior cervical ganglion and the superior thoracic ganglion of the sympathetic nerve trunk. The function of this trunk along with the rest of the autonomic nervous 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. Diseases of this trunk produce Homer's syndrome, characterized by vasodilation and increased blood flow. The same effect is achieved by anesthesizing the sympathetic nerve

trunk. Plester in sixties was the first to use this in treatment of sudden deafness. His results were encouraging which made many others to adopt to the similar line of treatment.

Neveling ( ) used the same and obtained good results. But his research an effectiveness of sympathetic chain blockage showed that blood volume decreased (and this was evidenced by increased nourishment of the cochlea.

A combined therapy of intravenous infusion of complamin and infiltration of stellate ganglion with 1% novacain was tried with various types of hearing losses by Sinzinger et al ('67)[21] . Based on the results the authors considered that this method was strongly indicated in cases of sudden deafness. A complete recovery which remained normal was recorded by Singleton ('71)[150]. Similar results have been reported by Haug('76)[158], Shaia et al ('76)[88] and Fujital et al ('79)[153]. Some of the complication of stellate ganglion block as put forward by Haug et al ('76)[158] are penumothorax, temporary loss of voice due to trauma ofthe recurrent laryngeal nerve and temporary dysphagia from trauma of vagus nerve.

Goodhill('72)[139] has given the management approach for patients with sudden deafness due to labyrinthine window rupture, which consists of use of such modalities like bed rest, intravenous procaine, histamine, vitamins, steroids and heparin singly or in combination. His own approach given in 1975 was to keep patient at absolute bed rest with head elevated 20° to 30°. There was no medication given other than an occasional sedative or tranquiliser. if no improvement occurred then a surgical intervention was done wherein perichondrium autografts were placed on the fistulous area. Post operative patients were kept on absolute bed rest with head elevated 20 to 30° for 48 hours. Later, he was restricted to sedentary activity for 5 to 10 days. Prophylactic antibodies usually ampicillin were prescribed for 10 days. He was restricted from flying, going to the mountains or strenuous physical activities for one month to several months. The above given approach was carried out in 15 cases and the results of the treatment are as riven in the table below:

No.	Age	Sex	Ear	Time of Surgery after sun-set.	Location of fistula	Pre operative SRT-SDS	post operative SRT- SDS
1	36	M	L	13 days	RW/OW	NR-NR	70- NR
2	57	M	L	13 days	OW	60-NR	10-80
3	45	F	L	1 month	OW	NR-NR	70-50
4	36	F	R	4½ months	OW	50-20	60-20
5	31	F	L	20 days	OW	NR-NR	75-NR
6	11	M	L	9 days	RW	NR-NR	65-70
7	29	M	L	4½ months	OW	62-30	80-16
8	58	M	L	2 years	OW	70-36	72-20
9	34	M	L	2 years	RW/OW	70-70	65-56
10	24	M	L	10 days	OW	90-NR	10-96
11	44	F	L	3 months	OW	NR-NR	NR-NR
12	52	M	L	9 days	OW	85-NR	0-76
13	51	F	L	4½ months	RW/OW	70-8	70-16
14	15	F	L	15 days	RW/OW	90-4	60-28
15	60	M	R	9 days	OW	82-24	70-24

Vestibulatory is one other surgical procedure which has been recommended to patients of sudden deafness due to labyrinthine window rupture by Fiedeman('75)[84].

The less popular of methods for treatment of sudden deafness are use of methylglucanine salts of which belong to the triohenaoic acid derivatives. (Fukuoka '78)[59], injection of amido-triovate-intravenously (Morimitsu ('74)[53], administration of hydrocortisone sodium succinate Ohtaf ('71)[16]. Microwave treatment was given by Kawamoto ('76)[67] who also reported excellent effects when care was taken to prevent over healing. The last of the treatment methods is use of ultrasonic's. (Kazama '76) .

C H A P T E R VIII

SUMMARY

In this project aa attempt has been made to compile the existing information on sudden deafness. The first chapter 'Introduction' gives a brief description of the nature of the problem. The definition of the condition as put forward by different authors have also been accumulated here.

In the second chapter which deals with etiology, the different conditions which have till now been illustrated ad leading to sudden deafness are described. Here it becomes clear that not only diseases or infections of the different parts of the ear and central nervous system, give rise to the condition, but also some of the surgical procedures can in turn be complicated, leading to the problem.

The third chapter on classification accumulates the classification system as put forward by various authors, which vary depending on the point of view from which it was derived. Eg. Some deal basically with etiology, while others are based on type of and extent of deafness.

The fourth chapter deals with pathogenesis and histopathology, which accumulates information on the changes that are seen in inner ear, as a consequence to the condition. This has got information got from experimental animals as well as those got from observation in human beings.

The fifth chapter deals with clinical signs and symptoms that are exhibited by these patients. There are a large number of features which extend from simple infections of the upper respiratory tract to severe vertigo with vestibular symptoms.

The sixth chapter deals with diagnosis of the problem which need results and investigation of not only the ear but all the systems of the body.

The seventh chapter deals with treatment of the problem. There have been a variety of methods given by different individuals. But till now there has been no one method that has been used with all and not all patients undergoing a particular treatment have had complete recovery. Besides this, a large number of patients also show spontaneous recovery thus making it difficult to decide whether or not a particular treatment is really appropriate.

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