SYSTEMIC DISEASE AND HEARING LOSS A REVIEW

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An independent project
submitted as part fulfillment of
First Year M.Sc., (Speech and Hearing),
Mysore.

ALL INDIA INSTITUTE OF SPEECH AND HEARING
Mysore

May,1998

Dedicated
to
Grand Father & Grand Mother
(MATHEW) (KAMALAM)

CERTIFICATE

This is to certify that this Independent Project entitled SYSTEMIC DISEASES AND HEARING LOSS A REVIEW is the bonafide work in part fulfillment for the degree of master of science (Speech and Hearing) of the student with Register No, M972L

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CERTIFICATE

This is to certify that this Independent Project entitled SYSTEMIC DISEASES AND HEARING LOSS A REVIEW has been prepared under my supervision and guidance.

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DECLARATION

This Independent Project entitled SYSTEMIC DISEASES AND HEARING LOSS A REVIEW is the result of my own study under the guidance of Dr. Rajalakshmi Lecturer in Audiology, Department of Audiology, All India Institute of Speech and Hearing, Mysore and has not been submitted earlier at any University for any other diploma or degree.

Reg. No. M9721

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May, 1998

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TABLE OF CONTENTS

INTRODUCTION

Hearing loss can be caused by multifactors. usually hearing loss in an individual occurs as an isolated entity. But often we do come across individuals demonstrating hearing loss with various other abnormalities.

Among this, systemic disease is one of the cause for hearing loss. Systematic Disease is one affecting a number of organs and tissues.

Dedicated investigators in the allied disciplines of psychology, speech pathology and audiology, pediatricians etc have contributed to the emergence of. various disease thus giving us a finer insight into the definite nature and cause of hearing loss.

The purpose of this project is to collect with in a single volume, the more important findings eminating from these diverse sources to enable the reader to get a comprehensive and varied knowledge about each diseases that is associated with hearing loss.

The information provided in the project will be of great help to researchers, teachers and future students in the field of speech and Hearing. This project contains chapters. Introduction, Review of Literature, (i) Systemic diseases (which causes hearing loss) summary & conclusion & Bibliography.

REVIEW OF LITERATURE

Crouzons' Disease

Premature synostosis of cranial sutures results in this distinctive autosomal dominant syndrome. The shape of the carniurn varies, depending on the sutures involved. Ocular hypertelorism, exophthalmos, shallow orbits. beaked nose, and maxillary hypoplasia are common. About one-third of the patients with this disease have a hearing loss (generally conductive) associated with ossicular deformity or stapes fixation. Bilateral external auditory canal atresias have been noted in some cases, as have mixed hearing losses. The treatment is surgical in most cases. Craniotomy is performed in infancy to decrease cerebral compression, with cosmetic maxillofacial reconstruction becoming more and more satisfactory.

Diabetes mellitus :

Diabetes mellitus is a chronic systemic disease related to a relative or absolute deficiency of insulin. It is classified as non-insulin dependent mellitus or insulin dependent diabetes mellitus which corresponds to the previous labels of adult onset diabetes mellitus (type II) and Juvenile onset diabetes mellitus (type I). They are suspected to be two separate entities from the pathogenesis with the non-insulin type probably the result of breakdown of interaction of regulatory mechanism. While insulin dependent diabetes, mellitus may be related to partial destruction of 'B' cells in the pancreas and may be modified by polygenic (suspected) to be a specific HLA antigen) and envisonmental (thought to be a viral infection) factors. An autoimmune component is thought to be responsible for the destruction of the B cells (Pickup and Willams, 1991).

Complications from insulin dependent diabetes are microvascular resulting in small vessel disease of the kidneys, retina and the skin as well as neuropathy, and less commonly macrovascular complications are seen : there is a higher risk of large vessel atherosclerosis, colonary disease and peripheral vascular disease.

Onset of diabetes is after 40 years of age in approximately 80% of individuals. In older individuals, diabetes is more prevalent in females than in males and hearing loss, if present, is characteristically bilateral. The onset of hearing loss is characteristically insidious, but may be instantaneous in a - few patients. Approximately 20% of patients will also complain of dizziness site of disorder is cochlea and/or eighth nerve degree of loss may vary from mild to profound and usually audiometric configuration will show greater loss in the high frequency region than in low or mid frequency regions.

Impedance results characteristically show normal, type. A tympanograms and normal static compliance measures. Acoustic reflex results vary, however. depending on the site of disorder and the degree of sensitivity loss. As a general rule, reflexes are present in ears with cochlear disorder and absent in ears on histologic examination, inner ear lesions are found in 50% of persons with diabetes (Torgensen and Buch, 1961).

A relationship between diabetes and sensoryneural hearing loss was first reported by Jordo in 1857. M. Kurien et al he found hearing threshold of 30 diabetic patients and 30 healthy controls attending the medical out patient department were determined using pure tone audiometry.

All subjects were less than 50 years old. Subjects with Otological and other metabolic diseases were excluded from the study. The patients were categorized into groups according to age, duration of disease. Complications and control of diabetes. These observations were compound with those from the control subjects using appropriate statistical methods.

it was found that diabetes had a poorer hearing threshold than the non-diabetics ; all age groups with diabetes showed a significant high frequency hearing loss, as compared to the control population : poorly controlled and complicated diabetics have significant, high frequency hearing loss as compound to those who were well controlled and uncomplicated ; there was no relationship between duration of the diabetes and the level *of* hearing loss. an excellent review by Taylor and Irwin (1978) endeavoured to nut this into perspective and made the following points from their own initial survey and from the literature. The incidence of senson neural hearing loss in diabetes will vary largely depend on the limits of normality and there in the statistical methodology second nearly all the work has naturally been carried out in the group of diabetics more likely to be affected, that is those on insulin. They were careful to limit their upper age range to 50 years, thereby reducing the effect of presbycusis. They noted that a diabetics with a family history had significantly better hearing thresholds than those without. They found that the diabetics, as a whole, were deafer particularly in the lower frequencies, than the controls and gradually approached each other in the middle range (1-4 KHz) and were similar at 8KHz.

Friedman and Schulman (1975) studied 20 diabetic patients with peripheral neuropathy ; 55% had a symmetrical hearing loss of the sensorineural type, involving at least one frequency, although none gave a history of hearing loss or ear disease. The hearing loss was unrelated to age and the impairment was similar at low and high frequencies, with maximum deficiency between 750 and 2000Kz.

K.V. Ravi, M.S., F.R.C.S., & A. Henderson, M.R.C.G.P. 1996 ; reported a 52 year old male with sudden loss of hearing in his right ear. With complaint of tinnitus in the right following decrease in hearing. There was no history of vertigo, earache, facial weakness, discharge, trauma, family history, viral infection etc.

An audiogram revealed 40dB to 70dB sensorineural hearing loss in the right ear. There was a 40dB hearing loss at 4000Hz in the left ear, possibly occupational. Tests for recruitment and tone decay indicated] a cochlear loss. They found increased blood sugar. The other investigations (routine medical checkup) were within normal limits after the insulin drip they found decrease in the blood sugar level (ie after treatment for 16 hrs.). The drip was discontinued and sugar level was controlled by diet alone. The patient reported definite improvement in his hearing thresholds.

Sieger et al (1983) a study in children but found statistically no differences in auditory function between insulin-dependent diabeties and normal controls or between diabetic with or without neurological or vascular complications. Brainstem responses also showed no difference between the two groups.

A small group of patients suffering idopathic sudden hearing loss was investigated by Wilson et al. (1982) to find out if there was a possible relationship to diabetes but no correlation could be found in the audiological pattern ; a similar incidence of recovery was noticed in the two groups through the middle frequencies ; however, the diabetic patients failed to recover as well in the high frequencies. Brain stem evoked responses also showed no abnormality and no evidence of retrocochlear dysfunction or pathology.

Mehra et al. (1985) investigated a series of 102 patients with diabetes and peripheral neuropathy to see if such patients were prone to dysfunction of the inner ear. Only 26 gave a history of hearing loss of mild degree, while 17 had tinnitus and 18 complained of vague giddiness. Investigations showed that one-half demonstrated some sensorineural hearing loss but when corrected for ageing, only 24 showed a mild loss (20-30dB). Eleven out of 91 showed markedly diminished caloric responses but all of these were in the older age group and had long-standing diabetes. Brainstem auditory evoked responses were carried out in 20 diabetic patients and a matched group of normal controls ; there was no difference in the latency of wave V and wave II although waves III, IV and V were delayed in the diabetic patients. Almost all studies on diabetic patients have been on those who are insulin-dependent.

A study by A. Parving, et al., 1990 ; they evaluated the cochlear and retrocochlear hearing function in patients with long and short term insulin dependent diabetes mellitus (IDDM) by means of Psychoacousic testing and auditory brain stem responses (ABR). Twenty patients with diabetic microangiopathy (median age 41 years, range 25-66 years) were examined.

The median duration of their diabetes was 26 years (range 13-46 years). In addition. 19 patients without micro angiopathy (median age 27 years range 17-42years) and with a median duration of the diabetes of 2 years (range 0-6 years) were examined. The metabolic control estimated by blood glucose concentration and glycosolated hemoglobin was identical in the two groups of **IDDM** patients. After correction for age and sex, no significant differences in hearing thresholds or discrimination scores were present between the two diabetic groups, or between the diabetic, patients and an age and sex matched background population. In the patients with long term IDDM, ABR produced abnormal responses in 40% indicating the presence of diabetic encephalopathy. whereas ABR were abnormal in only 5% of the patients with short-term IDDM.

Piras et al (1985) have reported a series of 30 diabetics of whom 27 were insulin dependent. Eight of the total group showed vascular lesions inherent to diabetics. They carried out auditory and vestibular studies both on the diabetic group and a similar number of normals controls and found that the influence of the disease was almost non-existent and the cochleo vestibular response was similar in both groups. Two centres in the UK (Nottingham and Cardiff) have combined in recent years. Gibbin and Davis (1981) investigated 50 diabetic subjects. 22 of whom were insulin dependent, the remainder being managed by other regimens and 50 control subjects. No significant differences were found between the two groups on pure tone audiometry or speech testing nor between those who were insulin dependent and those on different treatments.

Miller et al. (1983) from the same two centres investigated hearing loss in. patients with diabetic retinopathy. They found that the hearing threshold of patients with known diabetic retinopathy did not differ significantly from those of a control population. However, using a more suitable psychoacoustic test filtered speech task a definite difference in hearing acuity between the two groups was demonstrated.

Recent investigators have continued to find similar difficulties to their predecessors, some finding that there is no association between hearing loss and diabetes mellitus and others that there was quite definitely so depending at which sample you looked. Parving et al. (1990)investigated a group of long and short-term insulin dependent diabetics by a means of psychoacoustic testing and auditory brainstem responses. They tested those with and without diabetic microangiopathy. They could find no difference in the hearing threshold or discrimination scores between the two diabetic groups between the diabetic patients and an age and sex matched normal background population. Those with long term insulin dependent diabetes showed an auditory brain stem response abnormality in 40% with which they thought indicated the presence of diabetic encephalopathy. Whereas only 5% of those with short term insulin dependent diabetes showed an abnormal brain stem responses. Virtaniemi et al. (1994a) studied a series of 53 patients with insulin dependent diabetes mellitus and 42 randomly selected non-diabetic controls. They found that the hearing level tended to be worse in diabetic patients than in the controls but the difference was statistically different only in frequencies of 6000 Hz and 8000Hz, a finding of several previous studies. Macrovascular complications (retinopathy and nephropathy) and the duration of the diabetes was associated

with elevated hearing thresholds. By contrast, poor metabolic control was not associated with increased hearing thresholds. Virtaniemi et al. conclude that the elevated sensorineural hearing threshold at the two highest frequencies tested were probably caused by long duration of the diabetes and the microvascular complications associated it. The same authors (Virtaniemi et al., 1993) investigated auditory brain stem response latencies in insulin dependent diabetics and compared their findings with a degree of metabolic control, microangiopathy, neuropathy, and the duration of diabetics. In this study all the subjects had normal hearing ability. The wave V latencies were longer in the diabetic patients. Their overall findings seem to indicate a central disturbance of the auditory pathway and the microvascular complications and the duration of the diabetes were associated with prologation of auditory brain stem latencies. Poor metabolic control was only marginally associated with prolonged auditory brain stem latencies, but they felt that a causative role for diabetic neuropathy in the pathogenesis of prolonged auditory brain Stem latencies remained unresolved. The same authors (Virtaniemi et al., 1994b) also investigated the acoustic reflex response in patients with insulin dependent diabetes mellitus patients with diabetes had longer aconsic reflex latencies and decreased amplitudes compared with those of the control subjects. The acoustic reflex amplitude showed a linear correlation with the amplitude of the tympanogram, whereas the acoustic reflex latency had no linear correlation with auditory brain stem latencies in the same subjects. The acoustic reflex responses in the diabetic patients was not associated with the duration of the diabetes, is metabolic control, micro angiopathy or neuropathy. They felt the change was probably caused by stiffness in the middle ear system rather than disturbances in the brain stem.

Sigsbee W. Duck, M.D ; et al.,(1995) studied interaction between hypertension and diabetes mellitus in the pathogenesis of Sensorineural Hearing Loss".

The purpose of this study is to support the hypothesis that diabetic end organ damage of the cochlea is augmented in the setting of hypertension. A historical perceptive reviewing the effects of diabetes and hypertension as causative factors in the development of sensorineural hearing loss, as well as the basic epidemiology and pathophysiology of the renal and vascular effects Of diabetes and hypertension, is presented. The results of audiologic findings in insulin dependent diabetic patients, both normotensive and hypertensive. were analyzed and correlated with the results of animal studies to support the hypothesis that sensori neural hearing loss in patients and cochlear hair cell loss in animal studies result from the effect of hypertension in conjunction with insulin dependent diabetes mellitus.

Errors of Metabolism

Most of these disease are autosomal recessive. Tay-sachs disease, or amaurotic familial idiocy, is a sphingolipidosis caused by absence of heXosaminidase-A. This enzyme is now available for treatment of this disease. In addition, a blood test is available to detect carriers. In some areas, the test is performed routinely prior to marriage between Jewish people, among whom the disease is most common in this country. In the infantile form of the disease, flaccidity, motor regression, blindness with a macular cherry-red spot. and apathy begin at about 6 months of age. The disease progresses to spasticity andd death by age 3 or .4. In the juvenile form, loss of vision usually is the symptom; the course is slower and death usually occurs in the twenties.

High-frequency sensorineural hearing loss is felt to result from the basic metabolic anomaly. Gtitis media also is frequent, resulting in a possible mixed hearing loss.

Wilson's disease, or hepatolenticular degeneration, affects the brain, liver, and kidney and may result in deafness. The Kayser-Fleischer ring. involving Descemet's membrane of the cornea, is pathognomonic. The disease is caused by deficiency of plasma ceruloplasmin, the primary copper-containing plasma protein. This deficiency produces excess serum copper. The inheritance pattern is autosomal recessive, and a test to detect carriers is available. Treatment with fair results relies on low-copper diets and attempts to remove serum copper with chelating agents such as penicillamine dimercaprol (BAD and versene.

Fabry Anderson syndrome, a lipid-storage disease, produces mild sensorineural hearing loss is about 50% of patients. The disease is known also as cardiovasorenal syndrome of Ruiter-Pompen and diffuse angiokeratitis. it is characterized by elevated blood pressure, heart enlargement. angiokeratomata of the skin, pain in the extremities abnormalities of sweat secretion, and albuminuria. Frontal bossing and a prominent mandible and lips are common. Corneal clouding may occur, and macular purplish sports on lips and near skin mucosal junctions are common. Death frequently is caused by myocardial infarction or renal failure.

Other inborn errors of metabolism, including mannosidosis. other mucolipidoses, and other diseases, may be associated with hearing loss. Some

of these diseases may be relatively mild and are easily overlooked unless they are in the differential diagnosis.

Friedreich's ataxia

In Friedreich's ataxia there is no relationship between the progressive clinical involvement and the degenerative changes affecting the peripheral nerves. Pelosi et al. (1984) investigated a series of 15 patients of whom only five had a hearing difficulty (three mild one moderate and one severe). However brainstem electric responses were completely dissociated from the hearing disorder, being normal in one patient and abnormal in the remaining 12 investigated. Five showed severe abnormalities and there were mild to moderate abnormalities in the remaining seven, but wave I was present in all of this group. Patients without clinical acoustic disturbances showed abnormalities in brain stem response to the same degree or even greater than those who had a mild or moderate sensorineural hearing difficulty. However, the findings were significantly correlated with the level of clinical disability generally.

Visual evoked potentials showed abnormalities which corresponded to the severity of the clinical ophthalmological disturbance but were unrelated to the duration or severity of the clinical condition. Somrtosensory evoked potentials showed findings which were also unrelated to either the duration of severity of the clinical conditions.

Jabbari et al. (1983) studied five children in an effort to find out the primary site of auditory dysfunction in classic Friedreich's ataxia : none of

the children had any hearing complaints and all were tested soon after the onset of symptoms. The brain stem evoked potentials indicated dysfunction of the auditory system in the pontomesencephalic region. Acoustic reflex studies on two of the patients also suggested involvement of the brain stem auditory pathways. Wave I was retained in all patients and they thought it unlikely therefore that there was significant dysfunction of the spiral ganglia.

Ell, Prasher and Rudge (1984) reported on a group of 10 patients and noted that vestibular function and impaired hearing were common to most of the patients. Brain stem auditory evoked potentials were also abnormal in the majority. In their small series there was no obvious correlation between the type of severity of the neuro-otological abnormalities found and the duration of the disease. They commented on the high proportion of patients with vestibular and oculomotor abnormalities generally associated with cerebellar disease despite the absence of pathology at a gross level in that structure.

Hypoxia

Hypoxia or asphyxia during the perinatal period is thought to be a major causative factor in developmental problems in the neonate. But, although a history of hypoxia, or anoxia, is commonly elicited for hearing impaired infants, and McCormick and Davis (1994 personal communication) have suggested a definite association between recurrent apnoeic attacks and sensorineural hearing loss, it is now generally considered more likely to be one of a number of risk factors, the combination of which leads to damage. Good antenatal care and obsetetric parasctic minimize the incidence of hypoxia at delivery. These preventive elements, while commonly available in developed countries, are lacking in poor areas.

E.Borg done an overview of perinatal Asphyxia, Hypoxia Ischemia and Hearing loss. Birth hypoxia, asphyxia and ischemia have often been thought to be major causes of early hearing loss on deafness. The purpose of the present review is to focus on the role of these particular factors for perinatal auditory disorders. On the whole, only a small proportion of neonatal hearing loss is caused by perinatal factors. The exact etiology of, neonatal hearing loss in children with complicated deliveries is difficult to evaluate due to the large number of causative factors that might be involved. After reviewing the literature covering the past 15-20 years. It is not possible to say that we understand the relative importance of different factors and their inneractions. However, in the majority of studies, both asphyxia is not correlated with hearing Joss in babies with complicated deliveries. Prelonged artificial ventilation, the presence of severe hypoxic ischemic encephalopathy or persistent pulmonary hypertension are important factors. Tne brain is more susceptible to anoxia than the ear and both are more likely to be damaged after prolonged pre-peri- and postnatal hypoxia-ischemia than pure hypoxia during delivery. Perinatal hypoxia is more likely to cause a temporary hearing loss than a permanent one. Preterm babies are more vulnerable than term babies. The total number of risk factors, e.g. indicated by total length of stay in the neonatal intensive care unit and length of artificial ventilation, is the best predictor of risk for hearing loss of perinatal origin. The similarities between hearing loss and cerebral palsy are pointed out ; only *8%* of the cases of cerebral palsy are considered to be caused by conditions during delivery.

Hypothyroidism :

Schuknecht (1974) found the literature upto that time unconvincing on the relationship between acquired idiopathic hypothyroidism and sensorineural hearing loss, although commented that clinicians seemed to have the impression that there probably was such an association. Post (1964) investigated 42 patients seven with spontaneous primary hypothyroidism and 35 hypothyroid patients with treated carcinoma of the thyroid. He noted that slow mentation while hypothyroid may be interpreted by the patient as a subjective hearing loss. None of the patients with sensorineural loss attained entirely normal hearing when euthyroid. He was unable to demonstrate any specific correlation between age, degree of hypothyroidism and resulting deafness. He was also unable to determine the time required for patients to remain hypothyroid before experiencing a hearing less stephens and Hinchcliffe (1965) found a significant correlation between the diagnosis of myxoedima, and fatigue or temporary threshold drift measured at 8000Hz by the carhart technique. (Stephens 1970) later confirmed that this was not an artifaci relating to age. but a true finding. He suggested that the sensorineural lesion in myxoedema lies proximal to the hair cells. Meyerhoff (1976) reviewed the possible relationship between all forms of reduced thyroid function and hearing loss; under the heading 'non-genetic acquired' he reiterated the claims made up to that time, that is there was no definite association.

Van't Hoff and Stuart (1979) reported an incidence of deafness of 85% in a consecutive series of 48 patients with myxoedema. The more severe the disease, the higher was the incidence of deafness ; there was no difference between the effect on the high or low frequencies and in some cases the loss

was unilateral testing after the patients became euthyroid showed improved hearing in 73% ears. The percentage returning to normal (23%) showed no significant difference in the proportion of severe (20%) to mild myxoedema (26%). Repeat testing after becoming euthyroid, failed to show any further improvement. Age did not appear to be a factor in the cause of deafness in Myxoedeina. While severity of myxoedema was associated with a higher incidence of deafness. No other relationship could be found between seventy in myxoedema and a variety of other neurophysiological measurements. Van't Hoff and Stuart were in no doubt that the deafness was sensorineural.

Parving, Parving and Lyngsoe (1983) in a series of 15 patients with confirmed myxoedema, median age 76 years demonstrated a bilateral symmetrical or nearly symmetrical sensorineural hearing loss in all patients before treatment. Treatment with L-thyroxine in this group of elderly patients showed no improvement in hearing sensitivity and the group demonstrated neither more nor less hearing loss than other hearing - impaired patients *of* the same age group.

Hall et al. (1985) reported a prospective study undertaken to compare the auditory acuity in hypothyroid patients and to assess the effect of thyroxine on these thresholds, for a mean period of 5.7 months (range 2-24 months). Auditory thresholds were reduced overall frequencies but the difference being significant only at 2000 and 4000 Hz. Speech discrimination was also significantly reduced in both ears. With thyroxine there was a small improvement in pure tone thresholds and speech discrimination, this was only significant at 4000Hz in both ears.

Himelfarb et al. (1981) attempted to correlated changes in the brain item electric responses of patients with thyroid dysfunction six hyperthyroid : six hypothyroid). A good correlation was observed between the brain stem conduction time and level of serum hyperthyroidism, the brain stem conduction time was decreased and in some patients the brain stem electric response was characterized fey high amplitude waves, sharp peaks and fittery contours becoming smoother in pattern and more well-defined after treatment.

In untreated hypothyroidism, the brain stem electric response was generally characterized by prolonged conduction time, dimenished amplitudes, flattened peaks and poor synchronization ; in the older patients the changes in wave pattern were more pronounced. Brain stem conduction time appears to be a sensitive index of the thyroxine dependent cellular status in the neural pathways of the brainstem.

More recently, Anand et al. (1989) carried out auditory investigations in 20 patients with hypothyroidism they were assessed before and after treatment with L-thyroxine, 3-7 months after becoming euthyroid (mean 3 7) Sixteen demonstrated a hearing loss : mild in five, moderate in 11:12 sensorineural and four mixed special hearing tests revealed a cochlear type of hearing loss. Brain stem evoked responses showed prolonged, absolute latency of wave V and interpeak latencies I-IIl & I-V; the amplitude of waves I. 11 and V were reduced following treatment, a statistically improvement in the hearing threshold was observed by pure tone audiometry, but brainstem evoked responses did not show significant reversibility, subjectively nine patients complained of diminished hearing activity and three had tinnitus. Following

treatment, 13 patients reported hearing improvement, the hearing threshold improving by 5-10dB in eight and by more than 10dB in 12 at one or more frequencies, in one or both ears speech reception thresholds and speech discrimination scores were normal in all patients. Vanasse et al. (1989) investigated 15 adult hypothyroid patients by brainstem evoked responses before treatment and 17 also had pure tone audiometry. However, both tests were only repeated in five patients after treatment (2-22 months). They could find no significant changes in waves I, III, V latencies or in the I-V interval or in the I, III, V interpeak latencies, they commented on the different results achieved by themselves and Himelfarb et al. (1981), the latter authors measuring the brainstem conduction time from the peak of wave I to the peak of P4 (P4 being defined as the peak immediately following wave V and preceding wave VI).

Francois et al. (1993) investigated 11 congenital hypothyroid new born babies ; evoked otoacoustic emissions were present bilaterally in nine. Auditory brainstem responses in six new borns showed a prolonged wave I. When reassessed at 9-12 months of age after treatment, the auditory threshold was normal in nine and evoked otoacoustic emissions were present in at least one ear in all children. Four ears in two new borns could not be tested because the baby cried when the probe was inserted in the external ear canal. The authors found all waves on auditory brainstem responses to be clearly visible above the threshold in the congenitally hypothyroid new born although there it a prolonged latency of wave I there was no modification of the 1-1V interpeak latency. Earlier Hebert et al. (1986) using auditory brainstem response audiometry evaluated 34 congenital hypothyroid children receiving thyroid

hormone therapy ; their results suggested a significant incidence of auditory brain stem response abnormalities in treated hypothyroid children.

Parving et al. (1986) reported the audiological and temporal bone findings in Myxoedema. They investigated 15 patients with confirmed myxoedema with a median age of 48 years before and after treatment with L-hyroxine. No improvement in hearing sensitivity could be demonstrated either in the younger patients (age 32-60 years) or in the older group (64-95 years). When compared to an age and sex - matched in screened population. the myxodematous patients did not demonstrate any different degree of hearing loss. Histological investigation of the temporal bones from an 83-year-old woman with myxoedema, however, showed no morphological changes or deposition of glycosaminoglycans, changes which were compatible with true age related hearing loss. They concluded that those series which had previously indicated a hearing improvement after restoring the patients to a euthyroid state had been carried out on only a very limited number-of patients and when the sample was larger no abnormal hearing levels could be found either before or after treatment in myxoedema.

Hypoglycaemia :

Hypoglycaemia may be a factor in tinnitus that is worse immediately after sleep, especially on waking in the early morning. For such cases. R.R.A. coles advises the 'Lucozode test' (a glucose-rich mineral drink) in which the patient has half a cup ready to drink by his bedside ; when troubled by tinnitus on waking, the Lueozade is drunk with a little disturbance as possible. In about a quarter of such cases the tinnitus is reduced after 10-20 minutes.

either permitting a return to sleep or seeming to start off a day less troubled by tinnitus than usual. The notion of worse tinnitus being related to hypoglycaemia is supported by the author's experience of four insulin controlled diabetics whose most severe tinnitus occurred at times of impending hypoglycaemia and was immediately reducible by taking glucose.

Hypoglycaemia may cause acute dizziness and vertigo, and probably is the most frequent metabolic emergency (Binder and Bendtson, 1992). It usually results from inadvertent overmedication with insulin or oral hypoglycaemic agents. Much less common causes of hypoglycaemia include endocrine disorders, such as Addison's disease, hypopituitarism and rare insulin - producing tumours.

Hyperllpoproteinaemia {hyperlipidaemia)

This is one of the cause for fluctuating hearing loss. When considering this condition as a cause of fluctuating hearing loss, It is important to stress at the onset, the difference between primary and secondary hyperlipidaemia. There is a large number of conditions causing secondary hyperlipoproteinaemia, the most common of which are diabetes, alcoholism, chronic renal failure may also be a cause and oral contraceptives have been shown to elevate the plasma triglyceride in most subjects taking them. It is therefore essential to exclude these secondary causes, if not at the time of the original sampling, at least when the fasting lipids are being checked.

The next factor to be taken into account is the incidence of hyperlipoproteinaemia. Booth (1977) investigated 44 patients with premature

sensorineural hearing loss, without vertigo, and failed to find any incidence greater than in the local general population and no patient requiring treatment other than by a modification of the diet. Further cases have confirmed this finding and none so far has shown any significant improvement in hearing ; conversely there has also been no progression apart from age - related changes.

Drettner et al. (1975) in a study of 1000, 50 year old men investigated 8 number of cardiovascular risk factors to see if they might be of importance in the development of sensorineural hearing loss ; no significant correlations were found. Included among the risk factors, which were studied, were serum cholesterol, serum triglycerides, uric acid and glucose tolerance. Spencer (1981) has carried out the largest series associating abnormal lipids and inner ear symptoms. Of his 1419 patients, 18.4% were classified as having type 1IA or pure hyper cholesterolaemia with normal triglycerides ; 6.3% had type IIB primary hyper cholesterolaemia associated with lesser elevation of the cholesterol level. However, the incidence of obesity in these patients has varied from 72 to 100% depending upon the type of disorder and whether it was. associated with an elevated glucose tolerance. In his patients, reversing their dietary habits by cutting out refined carbonhydrates, reducing the intake of saturated fats and by increasing the amount of dietary fibre, avoidance of additional Salt and Sugar, and obtaining ideal body weight, he has reported improvements in hearing and has found similar therapy of value in treating patients with meniere's disease. Moffat, Booth and Morrison (1979) carried out detailed investigations including metabolic studies into 27 patients with Meniere's disease, but found no increased abnormality on glucose tolerance testing. fasting serum cholesterol and triglyceride levels, or estimations of

thyroid stimulating hormone. A similar evaluation was carried out by Kinney (1980) in 134 patients showing a high correlation of abnormal carbohydrate metabolism (5 hour test) and hyper lipoproteinaemia.

Lowry and Isaacson (1978) examined the audiology records of 100 patients presenting with a 20dB bilateral sensorineural hearing loss or greater. After a 14 hour fast and taking a history for the presence of diabetes the height, weight and blood pressure were recorded. Lipoproteins were estimated and 12 patients with type IIA or IIB and eight patients with type IV abnormalities were found ; they commented that such a finding was a lower proportion of patients with hyper lipoproteinaemia than in the general population. Jones and Davis (1992) investigated 279 men aged 50-60 years. Selected at random from those referred by general practitioners for lipid profiles. They found no relationship between the total cholesterol. Low density lipoprotein or triglyceride measurements and hearing loss could be found at any frequency upto 14KHz. The raised fasting glucose was associated with a higher hearing threshold at low frequencies ; a raised ESR was associated with higher hearing threshold levels in mid-frequencies. The effect of hish density lipoprotein level on hearing was highly dependent on the ESR. Vllrich. Aurbach and Drobik (1992) carried out a prospective study of hyperlipidaemia as a pathogenic factor in sudden hearing loss. They examined 25 patients with a first event of sudden hearing loss and nine with a repeated event of sudden hearing loss and nine with a repeated event of sudden deafness. Serum lipid patterns and atherogenic risk factors were the same in both groups and corresponded to lipid patterns in the average population histories of all the patients concerning smoking habits, alcohol and drug consumption.

noise, stress and recent viral infections were documented. They noted that flbrinogen is assumed to be much more critical concerning blood theology. but the plasma concentrations determined were with in the normal range in all patients.

Ben-david et al (1986) carried out a comparison of auditory brain stem evoked potentials in hyperlipid aemic and normolipaemic subjects. There were 25,hyperJipidaemic patients who were neurological and audiologically asymptomatic and 20 normolipoemic controls. Auditory brain stem evoked potentials showed the effect of increasing stimulus rate to be significantly greater in hyper lipidaemic patients (Click stimulus 10/s Versus (55/S). There were 17 type IV, one type IIA and seven type IIB in the hyperlipidaemic group. There was no significant difference in the brain stem auditory evoked potentials at a stimulus rate of 10/S. It was considered that the highly significant difference with click stimuli at 55/S indicated subcl.nical impairment of brain stem function in hyper lipidaemic subjects. Strome, tops and Vernick (1988) investigated the possibility of hyperlipidaemia being the cause of sensori neural hearing loss in children. They reported on this finding in three children two aged 6 and one 9 yrs of age but they give no indication as to how many others were tested and found to be normal/ negative.

Axelsson and Lindgren (1985) enquired about the possible relation ship between hyper cholesterolaemia and noise - induced hearing loss they compared 75 50 year old men randomly selected from the WHO material. The hearing was similar in both groups. They noted that noise was the most predominant factor influencing hearing at any specific frequency or combination

of frequencies. There Was a significant tendency for those with a high Cholesterol level who suffered the most noise exposure to have a high frequency hearing loss ; the tendency for the low cholesterol group was to high frequency hearing loss if excessively exposed to occupational noise.

Saadah (1993) reported on 31 patients seen over a 5-year period, mean age 58, in whom vestibular vertigo was associated with hyperlipidaemia and noted their response to antilipidaemic therapy. Complete resolution was finally achieved in 84% and it was noted that six of the 12 patients who stopped medication relapsed ; three of these who resumed therapy responded satisfactorily again. A cautionary reminder is given by Feher et al. (1992) emphasizing that in a group of 400 diabetic patients under their case despite regularly supervised diabetes, including dietary advice, over one-quarter had a raised serum total cholesterol (>6.5 mmol/1) while over one quarter of the non insulin treated and one eighth of the insulin treated diabetic subjects had a high density lipoprotein - cholesterol less than < 0.9 mmol/1.

Immuno Deficiency Syndromes :

Infection by the Human immuno deficiency virsus may affect the external. middle and inner ear in addition to the condition on occasion producing vertigo and facial nerve involvement. The overall picture is well summarized by Lalwani and Sooy (1992). Representative cases of the various presentations were published by Linstrom et al. (1993). The human immuno deficiency virus is a retrovirus with has the enzyme reverse transcriptase which functions to transcribe viral RNA to DNA, which is then incorporated into the genetic make up of target cell, the HIV virus has been shown to be

a lymphotropic virus, attacking principally 7-helper cells, as well as a neurotropic virus. As a result, AIDS is characterized by an underlying suppression of cell - mediated response leading to the development of apportunistic infections and /or malignant tumours. (Rarey, 1990).

In addition to the primary ear infection, secondary opportunistic infection is well recognized, the most common causative agent being toxoplasma, others being pneumocystis carinii and aspergillus; cryptococal meningitis may also occur [Breda et al., 1988; Kohan, Rothstein and Cohen, 1988; strauss and Fine, 1991; Hall and Farrior; 1993; Lyos et al., 1993}: Ofcourse mycotic infections of the temporal bone did occur before AIDS and even in the non-immuno compromised patient, but were rare (McGill, 1978; Meyer Hoff et al., 1979). Some 35-55% of patients with AIDS have a past history of venereal disease including syphilis, though some people with AIDS may not live longenough to develop to syphilis as it is a tertiary manifestation. The HIV virus may activate latent syphilis. It has also been noted that there is an increased incidence of oto siphilis and this appears to develop at an accelerated rate from the primary infections; it is though that this may occur because of the profound defects in cell - mediated immunity (Johns, Tierney and Felsenstein, 1987; Hart et al; 1989; Smith and Caralis (1980). In addition to the possible discharge from the external meatus and obvious hearing loss particularly due to conductive involvement, pans is frequently mentioned as ome of the presenting symptoms. Auditory evoked responses had been investigated in patients infected with the human immuno deficiency virus in an attempted to ascertain early brian stem involvement isolate changes with in the brain stem have been noted but. Birchall et al (1992) investigating

18HIV positive males in different stages found one-third had abnormal brianstem evoked responses or pure tone audio metry. However, they could find no correlation with the T-cell subset and only a week correlation with the pure tone audiometric average. Auditory and visual event related potentials were investigated by welkoborsky and Lowitzch (1992) and Baldeweg et al (1993).

Cases of sudden sensorineural hearing loss have been reported in HIV infection both as a presenting symptom (Timen and Walsh, 1989) with in a patient in whom the infection had already been diagnosed and with other symptoms evident (Real, Thomas and Gerwein, 1987). Grimaldi et al (1993) reported a patient who presented with a febrila illness and who. 2 days later. suffered a sudden bilateral hearing loss. Patients with an appropriate background who experienced a sudden bilateral hearing loss must therefore now be considered candidates for infection with human immuno deficiency virus intro other obvious cause in apparent.

Primary central nervous system lymphoma and systemic lymphoma with central nervous system involvement constitute the majority of central nervous system tumours in the AIDS population. Raposis sarcoma metastatic to the central nervous system is less common but may also be associated with hearing loss (Lalwani and Sooy, 1992).

In recent years temporal bone reports of ears involved by AIDS have been reported michaels, soncek with Liang (1994) reported on 49 bones from 25 patients, five with severe atitis media, 15 with low grade otitis media, two

with labyrinthine crypto coccosis, one kaposi's sarcoma with a deposit in the VIIIth nerve with six with cytomegalovirus in the inner and middle ear. Chandeasekhal. Siverls and sekhar (1992) also reported histo pathological and ultra structural changes but noted that the organ of corti was normal. Pappas et al. (1994) reported on the extra cellular viral - like particles with morphological characteristics of HIV/1 identified in the tectorial membrane in three cases morries and Prasad (1990) noted that the HIV virus, which is known to be neurotropic as well as lymphotopic, has never yet been cultured from the VIIIth cranial nerve or from spiral ganglion cells.

Klippel-Feil Syndrome

This well-known syndrome of cervical-vertebral fusion, often associated with spina biftda cervical ribs, neurological abnormalities strabismus, and other features, may be associated with hearing loss. When the Klippel-Feil anomalies are combined with abducens nerve paralysis, a retracted ocular bulb, and hearing loss, the condition is called Wildervanck's syndrome. A cleft palate or torticollis may be associated as well, and inheritance probably is multifactorial. Hearing loss may be unilateral or bilateral, moderate to serve. Both conductive and sensorineural deafness have been described, and vestibular studies are usually abnormal. Incomplete expression of the disease to common. The hearing loss is congenital and does not generally progress. Treatment may be surgical or may require a hearing aid.

Kartagener's Syndrome

The association of situs inversus, bronchiectasis, and sinusitis has been recognized as an entity since 1933 although earlier reports exist in the

liserature Inheritance probably is multifactorial, although an autosomal recessive pattern predominates. In addition to the classic findings poor pneumatization of the mastoid air cells and bilateral conductive hearing loss in the 30-to-dB range are common. The conductive hearing loss usually is due to middle-ear fluid, implicating eustachian tube obstruction. Middle-ear mucosal biopsy has shown chronic inflammatory changes. All patients with this disease need to be screened for hearing loss and to have their hearing restored medieaily, surgically, or, if needed, through use of a hearing aid.

Leopard Syndrome

Leopard is an acronym for: lentigines, electrocardiographic defect, ocular hypertelorism, pulmonary stenosis, abnormalities of genitalia. retardation of growth, and sensorineural deafness. The syndrome is transmitted as an autosomal dominant. The lentigines usually are absent at birth but develop progressively. Sensorineural hearing loss occurs in about 25% of cases and is usually mild. Treatment includes hearing aids where applicable, surgical correction of pulmonary stenosis, dermabrasion of the lentigines, and correction of other associated abnormalities as necessary.

Mallgraney

Primary carcinomas of sarcomas of the ear occur, but tumors form distant sites also may metastasize to the temporal bone. This fact is frequently unrecognised but should be considered if otitis develops in a patient with a known cancer. It also emphasizes the need to look for a primary tumor elsewhere when a carcinoma of the ear is found. Metastatic carcinoma to the ear has been reported from breast, kidney, lung, stomach, larynx, prostate.

thyroid. nasopharynx, uterus, meninges, scalp, rectum, the parotid gland, intestine, brain carotid chemodectoma, spinal cord, and other sues. Tumors of the skull base also can produce hearing loss by direct involvement of the ear or by interference with eustachian tube function, leading to fluid in the middle ear and conductive hearing loss. Direct extens.on from adjacent basal - cell earcinomas, melanomas, meningiomas, bengin or malignant neural tumors of nearby carnail nerves, glomus turmors hemangiomas (which may be multiple). and a variety of other neoplasms also may be implicated. Nasopharyngeal

cancers clssically occur with unilateral serous otitis media in an adult secondary to eustachian tube occlusions. This malignany is more common among Orientals but must be searched for in any patient with unexplained serous otitis.

Maliguancies such as Hodgkin's disease, leukemia, lymphoma. and myeloma,which produce defects in the immunologic system increase the incidenceof ear infection and resultant hearing loss and serious otologic complications. The importance of this possibility may be over looked in patients withdramatic systemic disease. Untreated otitis media can lead not only to a progressive hearing loss but also to meningitis and death, particulary in an immune - compromised patient.

Treatment for malignancy may involve radiation therapy. with its complications, dryness and scaling of the skin of the external auditory canal may lead to build up of debris and conductive hearing loss. Osteoradionecrosis

of the temporal bone may produce chronic infection and may result in

conductive or even severe sensorineural hearing deficit.

Meningitis :

Meningitis is the most common cause of severe acquired deafness in childhood, accounting for upto 90% of such acquired impairments in the UK. Incidence figures generally vary between b and 10% of the hearing impaired population, although Davis, in 1993, stated on incidence of 18% among profoundly deaf children with 12% among the severly hearing impaired and 3% of those with moderate losses. Fortnum (1992) quoted reports giving figures between 3.5 and 37.2% of those survivng meningitis having some degree of hearing impairmen. Immunization againstssome organisms which cause meningitis, is available.

Haemophillus meningitis :

Haemophillus influenzae is a Gram-negative coccobacilus and a major cause of infections in man. Invasive disease is associated with a high rate of morbidity and mortality and is caused by the encapsulated strains of the bacteria of which six types (a-f) are known to be pathogenic to man (Drugs and therapeutics Bulletin, 1993). Over 99% of invasive infections are due to type b (Hib) ; and affect 34 per 1,00,000 children per year. ie. one in 600 children will develop illness due to Hib before the age of 5 years. Sixty percent of these cases are meningitis, 15% epiglottis, and 10% septicaemia. H influenzae is responsible for 45% of all cases of meningitis. The mortality rate from meningitis is 4-5% with 11-30% developing permanent neurological sequelae such as sensorineural hearing loss. The disease has a high incidence between the ages of 3 months and 4 years, with a peak at 10-11 months of age. There had been an increase in cases in 1980s in th UK from 869 cases reported in 1983 to 1259 cases reported in 1989 (HMSO, 1992). The non-

encapsulated strains are associated with respiratory tract infection including otitis media.

Vaccines against H. influenzae were first introduced in the 1970s but were ineffective in very young children. Recent developments, where by the capsulae component, polyribosylribitol phosphate, is conjugated with either tetanus or diptheria toxoids, have increased their immunogenicity. Particularly in the very young. Animmunization programme was introduced in October 1992 in the UK where by Hib vaccine is given to infants as part of their primary immunization from the age of 2 months. The vaccine has previously been used in both the USA and finland and found to be very effective in reducing the incidence of invasive disease. Early reports indicate a marked reduction in cases of haemophilus meningitis in the UK since immunization.

Meningococcal meningitis :

Meningococcal meningitis accounts for up to 50% of cases of deafness occuring as a result of meningitis, although it causes only 15-25% of cases of bacterial meningitis Neisseria meningitidis or the meningococcus is a gram-negative diplococcus which gives rise to both meningitis and also meningococcal septicaemia, which can occur alone or in association. Asymptomatic nasopharyngeal carriage rates are around 10% (4-25% : Klein. Hey Derman and Levin, 1993) and spread is by droplets or direct contact from carries or individuals incubating the disease. The bacteria can be classified into antigenically different groups, of which A, B, C, Y and WI35 are the most common. Group B organisms are the major cause of disease in the UK. but group C organisms can be responsible for institutional out breaks. In

other parts of the world the risks of contacting meningococcal disease are relatively high with groups A and C causing many major epidemics. The aptly named 'meningitis belt' of Africa, lying between 15" and 5° North, being one such place.

The course of the disease can vary between being rapidly fatal within hours of the first symptoms-to a more insiduous and mildly progressive ulness. The maximum incidence is in infants and small children, with another peak of occurrence in teen agers and young adults mortality is quoted as being 10% ; 1-4% of cases suffer bilateral profound sensorineural hearing loss.

Vaccines are available against groups A, C. Y and WI35 and are used to combat small out breaks as well as more widespread epidemics. There is no,vaccine available for group B meningococcus, and instead, prophylaxis (oral rifampicin) for contacts of a confirmed case is recommended with vaccination only if the ulness is caused by group C (or rarely group A) bacteria in the epidemic areas mass vaccination is considered to be of value. although reports suggest that sero conversion in children is low (Klein. Hyberman and Levin, 1993) and longterm protection is poor (Ceesay et al.. 1993). There is a need for more research into long term effective vaccines against all strains of the disease.

Pneumococcal meningitis :

There is no effective vaccine against pneumoccal meningitis, although vaccines have been developed to combat other pneumococcal diseases.

Steptococcus pneumoniae is an encapsulated Gram positive coccus. There are about 84 recognized strains of which 23 types cause 90% of cases. Infections with pneumococcus are relatively common and carey a high mortality and morbidity rate. The pneumococcus is responsible for about one fifth of the cases of bacterial meningitis, with a mortality rate of 50% and with 25% or more of survivors having residual neurological problems of which sensorineural hg loss is common. The vaccine currently available is against the 23 commonly pathogenic capsular types of pneumococcus and is reported as being 60-70% effective is preventing pneumococcal pneumonia consequently it is of particular value post splenectomy. It's efficacy is children is reduced, particularly in those under 2 years of age. and it does not prevent otitis media nor exacerbations of chronic bronchitis. As there primary infections lead on to pneumococcal meningitis this vaccine therefore has little value as a preventive agent against meningitis.

Research is continuing into vaccines against this disease.

Incidence :

The most frequent cause of acquired sensonneural deafness in children (Martin,. 1982) Davis and Wood, 1992). The latter estimated that about 90% of acquired deafness was due to this cause. Fortnum (1992) reviewed the literature. The incidence of post-meningitis hearing impairment is reported as varying from 3.5% to as high as 37.2%. This large range reflects the sampling errors associated with small samples, the type and seventy of the hearing impairment included, the timing of the assessment after the onset of the cases,the age range of the children, the profile of the infecting organisms.

the sophistication of the tests used and the reliability of the referral system. In Belfast it was found that only about 50% of children with bacterial meningitis were referred for hearing assessment. Subsequently a protocol has been devised to ensure that all children have their hearing assessed with in 1 month of discharge from hospital. It would appear that the true incidence of sensorineural hearing loss after meningitis lies between 8 and \2% with about one quarter of these having profound sensorineural deafness.

Fortum and Davis (1993) considered the risk factors for Sensorineural hearing loss in children with meningitis. These included the following the presence of associated hydrocephalus, children aged less than 1 month or greater than 5 years, children whose length of stay was greater than 16 days in the hospital, admission during the winter month (October-March), and a cerebro spinal fluid (CSF) glucose concentration of less than 2-2 mmol/1. It would appear that there is no differential risk for the infecting organisms.

The ear may be affected in different ways by meningitis. Bacterial labysinthitis due to direct spread of the injection from the subarachvoid space through the cochlear aqueduct, internal acoustic meatus or endolymphatic duct is associated with profound sensorineural hearing loss. In children who have portial or reversible hearing loss there may be toxic or serous labysinthitis.

The deafness is usually bilateral and profound although may be less severe and even unilateral (Rasmussen, Johnsen and Bohr, 1991). There have been reports of improvements in the hearing after meningitis even in children with profound bilateral sensoreneural hearing loss, although improvements are

more often reported in cases with less severe problems. Fortran considered that some of these may have been due to improvements is the reliability and accuracy of hearing tests as the child got older or to resolution of a simultaneously present conductive hearing loss due to fluid in the middle ear. Kulahli, M.D., et al 1997; evaluated in early and late period of bacterial memngitis children with auditory brain stem responses.

The hearing function of 50 children with bacterial meningitis was evaluated at the second and 10the days, and eighth weeks after admission with auditory brain system responses (ABR) to investigate whether meningitis causes hearing loss normal values were obtained in all tests from both ears of 24 patients (48%). Twelve patients had temporary, and seven patient had persistent mild degree hearing loss severe hearing loss was detected bilaterally in five patients and unilaterally in two patients patients with other complications such as subdural effusion, convulsion, brain oedema and paralysis were found to have a higher incidence of hearing loss. We observed that patients treated with dexametharome had 7.7% persistent hearing loss 11.6% mild hearing loss 34.6% transient hg loss, but in the group who did not receive dexametharome there was 19.2% persistent hg loss 15.3% mild hg less and 11.6 percent transient hg loss. There were other significant differences between the two groups in restoration of normal body temperature the CSF/plasma glucose concentration ratio was evaluated CSF (uretro-spinal fluid) protein concentration was decreased and the cell count in the CSF was decreased in the dexametharone group, significantly more than the group who were not receiving dexametharone. The hearing loss tended to be more frequent among younger children.

Fraxin Robert Amaee et al., 1997 studied possible involvement of nitric oxide is the sensorineural hearing loss of Bacterial meningitis.

Micro perfusion of scala tympani with the no donors, sodium nitroprusside (SNP) and S-nitroso-N-acetylpenicillamine (SNAP), Produced marked depression of the compound action potential (CAP) and cochlear Microphonic (CM) together with severe and widespread morphological damage to hair cells and supporting cells the organ of corti. In addition, direct perfusion of N-methyl-D-aspartate (NMDA) into scalatypani, which probably induces excess stimulation of NMDA receptors with in the cochea and which is known to elad to the release of NO, was found to elicit similar electrophysiological and structural lesions in the cochlea. Pre-perfusion of Scale tympani with L-methyl orginine (L-MA), which inhibits the release of No, or super oxide dismutase (SOD), an O_2 scavanger, conferred marked protection upon the cochlea from the lesions caused by No donors. These observation indicate that enhanced No production is likley to be an important factor responsible for pathological insult of the chchea. The possibility is discussed that this factor is involved in the chain of events leading to hearing loss caused by bacterial meningitis. Such hearing loss is a major sequela of bacterial meningitis in children.

The common organisms associated with sensorineural-hearing loss after meningitis are stereptococcus pheumoniae, haemophilus, influenzae and Neisseria meningitidis. In spite of previous publication suggesting that Neissena meningitidis is the most dangerous organism with respect hearing loss, it would appear that bacterial meningitis of any type can result in sensorineural

hearing loss of any degree in a child of any age and that meningitis sensorinev hearing loss is not truely organism specific (Fortmum, 1992).

Meniere's disease :

Incidence :

The estimates of the incidence of Meniere's disease in the world literature vary significantly, In.Great Britain, Cawthorne and Heweltt (1954) reported an incidence of 1 per 636 persons per year, or 157 per 1.00.000 population. Harrison and Naftalin (1968) reported an incidence in the UK of 0.1% or 100 per 1,00,000. In Sweden, in a study examining hospital discharge and outpatient data on a population of over two million, the incidence of Meniere's disease was found to be 46 per 1,00,000 (Stahle, Stahle and Arenberg, 1978). In France, Michel, Fouillet and Trovero (1977) have indicated an incidence of only 7.5 per 1,00,000. In Japan, the incidence of Meniere's disease appears to have increased dramatically since the Second World War (Watanabe, 1981). In Africa, the diagnosis of Meniere's disease is uncommon. despite the fact that vertigo is a common presenting complaint (Watanabe. 1983). Estimates of the prevalence of Meniere's disease are even more difficult to obtain than incidence figures. In view of its nature as a non-lethal non-communicable disease, inadequate public health records exist in most countries. Based on the Swedish study, however, conservative estimates and data indicate that Meniere's disease is at least four times more common than clinical otoscleosis (Arenberg et al., 1980). Clinically, Meniere's disease is responsible for 10% of visits to a busy dizziness unit in a tertiary referral centre (Nedzelski. Barber and Mcllmoyl, 1986). Meniere's disease is rare in the south western American Indian population (Pfalz and Thmosen, 1986).

A sexual preponderance in Meniere's disease has been suggested by the work of some authors. In Sweden, Stahle, Stahle and Arenberg (1978) found a female preponderance in a ratio of 3:2. In Japan, a 3:2 male preponderance in a data survey from 1934 to 1960 disappeared when the survey was more rigorously repeated in 1981 by Watanabe. Balkany, Pillsbury and Arenberg (1980) found no sexual preponderance, nor did Paparella (1985). The right and left ears are affected with equal frequency.

A familial tendency has been described in Meniere's disease, with a positive family history in up to 20% (Paparella, 1985). Birgerson, Gustavson and Stahle (1987) found that 14% of affected patients had a first order relative with the disorder, including a family with three generations of Meniere's disease and another with an aberration in chromosome 7. It is generally felt that, while genetic transmission may play a role in Meniere's disease, transmission is variable and inheritance is multifactorial (Paparella 1985).

The age at onset of symptoms is extremely variable. Meniere's disease in children under the age of 10 years is rare, with the youngest case described in a four year old. The disease begins in most patients before the age of 60 years (Thomas and Harrison, 1971; Greven and Oosterveld, 1975; Stahle, Stahle and Arenberg 1978) with a noticeable peak in incidence in the fifth and sixth decades of life (Oosterveld, 1980).

The frequency of bilateral Meniere's disease ranges from 2 to 78% in the literature (Balkany, sizes and Arenberg, 1980; Green, Blum and Harner.

1991). The difficulties that may give rise to this wide range are the lack of consensus about the diagnostic certeria and the length of time Meniere's disease patients should be followed up for the development of contralateral disease. In the largest study, a British survey of 610 patients with Meniere's disease followed for at least 5 years found that the incidence of bilateral disease was 31.8% (Thomas and Harrison, 1971). Friberg, Stahle and Svedberg (1984) reported an incidence of 47% in 34 patients who were followed for 20 years, and showed a relationship between incidence and follow-up observation time. Kitahara (1991) found a 9% incidence in the first year of follow up and 41% when the patients had been observed for more than 20 years. It is now generally agreed that the incidence of bilateral disease increases continuously over time (Paparella and Griebie, 1984; Stahle 1991). Subclinical Meniere's disease may exist in the second ear long before the development of overt symptoms (Moffat et al., 1992). Greven and Oosterveld (1975) found 10% of 292 patients had developed classical Meniere's disease in the second ear, but that 73% of the patients in this group had signs of hearing disturbance in the second ear, such as sensorineural hearing loss, recruitment, or tinnitus.. Paparella and Griebie (1984) studies 360 patients the same way and obtained numbers of 32% and 78.6% respectively. More compelling are recent results using transtympanic electrocochleography, the only proven investigation that can demonstrate objectively the presence of endolymphatic hydrops. Electrophysiological evidence of hydrops was seen in 35% of second ears in a population of 40 patients with unilateral clinical Meniere's disease (Moffat el al., 1992). The early recognition of incipient Meniere's disease in the asymptomatic contralateral ear of the patient with known unilateral disease has profound implications for patient management and follow up. The high

frequency of bilateral disease dictates that, when considering therapeutic options, every attempt should be made to conserve hearing.

Pathophysiology

Meniere's disease appears to be one member of a group of disorders of the inner ear linked by the common pathophysiological condition of endolymphatic hydrops (Hallpike and Cairns, 1938; Altmann and Kornfeld. 1965; Antoli-Candela, 1976; Schuknecht and Igarashi. 1986). Endolymphatic hydrops is thought to be a pathological condition that is the, end result of a variety of insults to the inner ear, and may be subdivided into symptomatic and asymptomatic forms. This was further subclassified by Schuknecht and Gulya in 1983. The symptomatic form is characterized by the classical triad of fluctuating sensorineural hearing loss, episodic vertigo and, usually, tinnitus and the asymptomatic form is clinically silent (Rauch, Merchant and Tuedinger, 1989). The acute attacks of the symptomatic form are superimposed on a gradual progressive reduction in the auditory and vestibular functions of the affected ear over time.

Endolymphatic hydrops is a physical distortion in the membranous labyrinth. Since the original report of endolymphatic hydrops by Hallpike and Cairns (1938) and Yamakawa (1938), 134 temporal bone cases have been reported (Paparella, 1985). Cochlear hydrops was seen in all cases and saccular hydrops was seen in most. Utricular hydrops was uncommon. Endolymphatichydrops therefore was observed most consistently in the pars inferior (Altmann and Kornfeld, 1965; Schuknecht and Igarashi, 1986; Okuno and Sando, 1987) and could be identified by the typical bowing of Reissner's

membrane and distension of the saccule (Schuknecht, 1974), Enlargement of the endolyphatic space clearly occured at the expense of the perilymphatic space (Antoli-Candela, 1976; Klis, Buijs and Smoorenburg. 1990). The degree of endolymphatic space expansion was variable. The endolymphatic space bulged in the region of the helicotrema in half of the cases, exhile the saccule bulged against the footplate in 60% of the cases reviewed by Paparella (1985) arid into a semicircular canal (usually the horizontal semicircular canal) in about one-third of cases. Fibrous adhesions can form between the saccule and the under surface of the stapedial footplate (lssa et al., 1983). This contact may explain Hennebert's sing, which is subjective vertigo. and tonic eye deviation and nystagmus observed during a pressure-induced excursion of the footplate (Nadol, 1977). It may also explain the Tullio phenomenon which is experienced by some Meniere's patients, namely a subjective imbalance and nystagmus observed in response to loud, low frequency noise exposure (Tullio, 1938; Ishizaki et al., 1991).

Changes in the pars superior (utricle and semicircular canals) are observed less frequently and are less dramatic, and more likely to be seen in cases of longstanding Meniere's disease. Utricular dilation and herniation into the crus commune has been observed (Lindsay, 1942), as well as ampullary distortion and displacement of the cupula from the ampullary wall (Antoli-Candela, 1976; Rizvi, 1986).

Interestingly, despite the progressive decline in auditory and vestibular function witnessed in these patients over the years, there is relative sparing of hair cells and the first order neurons in this disease. Only in the most

severe cases will these structures show damage and a depletion in numbers (Schuknecht and Igarashi, 1986; Schuknecht, Suzuka and Zimmerman, 1990) Endolymph is derived predominantly from the stria vascclaris; the planum semilunatum and dark vestibulars; the planum semilunatum and dark vestibular cells contribute a small amount. Endolymph may also be produced from perilymph across the labyrinthine membranes (Paparella, 1980). The circulation of endolymph is both radial and longitudinal (Lawrence, 1980). The longitudinal pattern starts with the production of endolymph in the stria vascularis of the cochlea, circulation via the scala media occurs through the ductus reuniens to the saccular duct, where it proceeds into the vestibular labyrinth. Elimination of endolymph occurs via circulation through the vestibular aqueduct and on to the endolymphatic sac, where it is absorbed Radial flow results from the production of endolymph in the dark vestibular cells and planum semilunatum with local absorption. The evidence strongly suggests that both longitudinal (slow process) and radial (rapid process) circulations are concurrently operational and subject to both hydrostatic and osmotic pressure gradients.

Endolymphatic hydrops occurs through the accumulation of endolymph. either through its overproduction (Henriksson, Gleissner and Johansson. 1986 or through its inadequate absorption. The prevalent theory is that the fundamental problem is one of longitudinal flow, specifically endolymphatic malabsorption, with the site of this dysfunction being the endolymphatic sac or duct (Schuknecht, 1968; Paparella, 1985). This concept is supported by several studies in which endolymphatic hydrops has been induced by injury and disruption of the endolymphatic sac in guinea-pigs, rabbits and cats. The

successful mechanisms of this injury include mechanical means (Kimura and Schuknecht, 1965; Beal, 1968; Horner, Erre and Cazals, 1989). Chemical cauterization (Yazawa, Shea and Kitahara, 1985), viral inoculation and infection (Fukuda, Keithley and Harris, 1988), or by an immunologically induced inflammatory response (Yoo, 1984; Sawada et al., 1987; Tomiyama. 1992). Despite this, the hydrops which is histologically identified in these animal models has not resulted in the clinical presentation of the symptoms of Meniere's disease experienced in humans.

Even in the normal ear there are enormous variations in the surgical anatomy of the endolymphatic sac (Friberg et al. 1988. Studies in humans with endolymphatic hydrops have revealed hapoplasia of the vestihular aqueduct (Sando and Ikeda, 1984), narrowing of the endolymphatic duct (Yuen and Schuknecth, 1972; Ikeda and Sando, 1984), perisaccular fibrosis (Altmann and Fowler, 1943), loss of epithelial integrity and atrophy of the sac (Arenberg. Marovitz and Shambaugh, 1970), and positive immunofluorescent staining for immunoglobulins of the sac wall (Yazawa and Kitahara 1989). Temporal bone studies of otosclerosis have implicated bony narrowing of the vestibuiar aqueduct and duct obstruction in explaining the coexistence of otosclerosis and endolymphatic hydrops (Sismanis, Hughes and Abedi, 1986; Franklin. Pollak and Fisch, 1990; Yoon, Paparella and Schachern, 1990). The wall of the endolymphatic sac in Meniere's disease has been shown to have significantly fewer and smaller blood vessels than normal controls, suggesting a microvascular contribution to the pathogenesis of the disorder (Ikeda and Sando, 1985). When the lumen of the endolymphatic sac is studied in patients with Meniere's disease, stainable proteinaceous material can be seen (Takumida.

Bagger-Sioback and Rask-Anderson, 1989; Friberg et al., 1988). Other authors have found this secretion to have the staining characteristics of a glycoprotein and have suggested that the endolymphatic duct may function as an organ of merocrine secretion as well as endolymph absorption (Wackym et al.. 1990; Rask-Anderson et al., 1991). Wackym (1995) has recently suggested three possible pathophysiological mechanisms to account for the histopathological changes in Meniere's disease; fibrosis of the endolymphatic sac and vestibular epithelia, altered glycoprotein metabolism and inner ear viral infection. Tightly adherent dura in the region of the endolymphatic sac has been seen at the time of revision endolymphatic sac surgery, and this is thought greatly to increase local sac pressure and resistance to endolymph flow, supporting the malabsorption theory (Paparella and Sajjiadi, 1987a).

Blockage of the duct and hypoplasia of the sac, however, are not seen in all cases of Meniere's disease, and it therefore seems likely that other mechanisms may play a role in the development of endolymphatic hydrops. Stahle an Wilbrand (1983) described a lack of periaqueductal pneumatization. with its concomitant effect on the angulation of the vestibular aquedut within the inner ear, lack of pneumatization medial to the arcuate eminence, a short vestibular aqueduct with a narrow external aperture and a reduction in the overall size of the mastoid air cellsystem as the characteristic features of the temporal bones of patients with Meniere's disease which they studied. Sando and Ikeda (1985) also noted the poor mastoid pneumatization, although others have been unable to verify this finding (Arnhold-Schneider, 1990). Paparella et al. (1989) noted a significant anterior and medial displacement of the lateral sinus and consequent reduction in the size of Trautmann's triangle in

patients with Meniere's disease. They hypothesized that long-term endolymphatic malabsorption is related to a developmental abnormality of the endolymphatic duct and sac in association with hypogenesis of Trautmann's tringle. Impeded local venous drainge, such as one might see in a displaced lateral venous sinus, may result in a disruption of hydrodynamic forces in the labyrinth and endolymphatic hydrops (Gussen, 1982).

The histopathological discovery of membrane ruptures and their repair is compelling evidence for this theory and moreover, Tasaki and Fernadez (1952) found that perfusion of the perilymphatic compartment of the cochlea with a solution of potassium blocked the cochlear microphonic responses and action potentials. This would explain the difficulty in recording these response when transtympanic electrocochleography is carried out in a patient during an acute attack. A similar acute, intense, reversible paralysis of vestibular function was shown when the perilymphatic space was perfused with artificial endolymph (dohlman and Johnson 1965; Silverstein, 1970). Other groups have detected changes in the composition of endolymph after several weeks of experimentally-induced endolymphatic hydrops. suggesting that hydrops itself may cause pathological permeability of the endolymph-perilymph barrier (Jahnke, 1981; Szikalai et al., 1989).

Despite all of this evidence, the theory remains controversial, however. as some authors believe that membrane ruptures occur rarely and constitute catastrophic events in the inner ear (Tonndorf, 1968, 1986; Thomsen and Brctlau, 1986). A belief held by some is that high endolymphatic pressure alone can produce the Meniere's symptom complex. Animal work by Andrews.

Bohmer and Hoffman (1991) has looked at the pressure measurements of endolymph and perilymph in normal and hydropic ears and found that endolymph pressures in hydrops are elevated compared with normal endolymph and perlymph pressures. The cochlear damage and hearing loss of Meniere's disease can be reproduced by manipulating and increasing endolymphatic pressure alone (simmons and Mongeon, 1967). Paparella (1985) has pointed out that the saccule blocks and fills the vestibule, and that a break in the membrane of the cochlea may not therefore result in contamination of the perilymph of the pars superior. He has reasoned that multiple membrane ruptures would therefore be necessary to produce the classical symptoms of Meniere's disease, an unlikely event which is not supported by existing temporal bone studies. He has proposed that the observed ruptures actually alleviate the sumptoms of a Meniere's attack, a theory supported by the fact that many patients enjoy relief from the attack when the aural pressue subsides.

Mucopolysaccharidoses :

Clasically, six varieties of inborn metabolic errors involving mucopolysaccharide metabolism can be described. Hurler's syndrome (gargorlism) is characterized fades, claw like hand deformities, and death before the age of 10 years. The condition usually is recognized during first year of life and the inheritance is autosomal recessive. The nasopharynx is deformed and lymphoid tissue is markedly increased, leading to nasal obstruction and chronic nosal discharge. This discharge may cause eustachian tube obstruction, compounding middle-ear disease. Primary pathology also be found with in the middle ear, however, apparently resulting from the presence of the disease in utero. Hurler's syndrome can be diagnosed by amino centesis.

Sensori neural hearing loss may occur in this disease, but it is usually mild. Hunter's syndrome has a similar habitus but does not show corneal clouding. It is X-linked, so the disease is expressed only in males. The onset of signs

and symptoms usually occurs around age 2, and most patients die by the age

of 20. However, some have lived into their sixties. About 50% of these cases

are accompanied by progressive hearing loss. The loss usually is not serve

and is most commonly mixed or sensorimeural.

In sanfilippo's syndrome, an autosomal recessive anomaly, hearing loss is uncommon. When present, it appears around the age of 6 or 7, and then it progresses. These patients live a nearly normal life span and develop their symptoms early in childhood. They show progressive mental deterioration. mild coarsening of the facial features and stiffening of joints.

In Morquio's syndrome, also autosomal recessive, mixed hearing loss is common and usually begins in the teen years. Onset of symptoms is . between ages 1 and 3, and the features are very similar to those of Hurler's syndrome, including the corneal clouding. The coarsening of facial features is milder, however, severe kyphosis and knock knees are characteristic!

Scheie's syndrome is an allelic form of Hurler's syndrome. The broad mouth and full lips characteristic of gargoylism are present by the age of 8 years. Corneal clouding occurs, as do retinal pigmentation, hirsutism and aortic valvulal defects, is also may be found in the other mucopolysaccharidoses. Psychoses and mental retardation may occur but are not as striking as in the related syndromes. The life-span is long and the

inheritance, is autosomal recessive. Although documentation is inadequate. it is suspected that mixed hg loss develops in 10-20% of these pallets, usually in middle age.

Patients with maroteaux - Lamysyndrome show features similar to those of Hurler's symptoms and signs some what later than in Hurler's syndrome. and deformities are generally less severe.. By about the age of 8 years, roughly 25% will exhibit hearing loss (probably conducting apparently associated with recurrent otitis media.

Diagnosis of the mucopolysaccharidoses is confirmed by detecting stored or exerted specific mucopolysaccharides. As yet, treatment is not available for most of these diseases. However, in Hurler's syndrome, the enzyme α-L-iduronidase is now being used with some favaourable results.

Pseudo-Hurler's syndrome is an autosomal recessive generalized gangliosidosis, rather than a mucopolysaccharidosis. The defects are severe. similar to those of Hurler's syndrome, and they include a cherry-red spot in the macula of the eye in about half of the patients. Death generally occurs by 2 years of age. Information regarding hearing loss is lacking and will not be truely relevant until some effective therapy is found for the underlying disease.

Deafness is a well-recognized components of the clinical phenotype in both Hurler's and Hunters diseases (Hurler, 1919 ; Kittel, 1963). Although both these diseases are invariably fatal, deafness often makes an appreciable

contribution to the overall morbidity in the earlier stages of their evolution. Hearing loss is also an important practical problem in the clinically milder symptoms associated with α-L-iduronidase deficiency, scheie disease (MPS.1S) and Hurler scheie disease (MPS IH/S), and in the mild variants of Hunter's disease Briedenkamp et al. (1992) in a comprehensive review of 45 children with mucopolysaccharidoses have determined the frequency of complications related to the head and neck. Recurrent respiratory infections occurred in *3%* and chronic recurrent middle ear infections with effusions occurred in 73% of the patients. The significance of these complications lies in the fact that their continuing care is often 'the primary management issue of these patients' (Briedenkamp et al., 1992). All cases of otitis media but one responded to antibiotic treatment and dry ear precautions. One patient required mastoidectomy and there was a great deal of granulation tissue present filling the middle ear. Histological examination of this tissue showed the typical foamy histiocytes with deposits of intracellular mucopolysaccharides as described in our cases of Hurler's disease (Friedmann et al.. 1985). Sensorineural hearing loss was observed in four patients.

Histopathology :

Characteristic vacuolated Hurler or gargoyle cells were noted disrupting the fascicles of the vest ibulocochlear nerve within the temporal bone in two cases of Hurler's disease (Schaechern, Shea and Paparella, 1984 ; Friedmann et al., 1985), the perivascular spaces of the mastoid process contained many vacuolated cells, and large areas of the mastoid process were replaced by accumulated Hurler cells. The neuro radiological features in these cases were described by Watts et al. (1981) and postmortem bio chemical and general pathological studies were reported by Crow et al. (1983).

Khetarpal et al. (1991) have described in five temporal bones from three subjects of two kindreds with an inherited form of sensorineural hearing loss (autosomal dominant), the accumulation of an acid mucopolysaccharide with in the bony channels of the dendrites of the VIIIth nerve causing degeneration of neural and sensory structures. All of these patients demonstrated a progressive high frequency sensorineural hearing loss since early adulthood but only one had a history of recurrent vertigo. However, all the temporal bones showed consistent pathological findings of the mucopoly saccharide material obliterating the bony channels of the vestibular and cochlear innervation pathway. Based on this series as well as previous reported temporal bones of this form of sensorineural hearing loss, the authors proposed two phenotypes of autosomal dominant hearing loss :

1. The progressive sensorineural hearing loss with an onset in the third to the fifth decades having the appropriate pathological correlates of sensory and neural degeneration in the organ of corti but with a normal vestibular labyrinth. Vestibular symptoms are absent in this phenotype.

2. The sensory and neural degeneration is present in both the auditory and vestibular labyrinth with the mucopolysaccharide material obliterating the bony channels in the cribrose areas of the labyrinth producing retrograde degeneration of both the vestibular and auditory neurons. This accumulation of mucopoly saccharide is also responsible for degeneration of hair cells and supporting cells of end organs by infiltration of the stroma of the sense organs. The proposed mechanisms by with the mucopolysaccharide material may cause neural and sensory degeneration is by compression of the dendritic processes of the neurons, alteration.

of the fluid environment of the neurons or compromise of the blood supply to the neurons and sense organs. It is quite possible that a combination of all three mechanisms may be present in this particular form of hereditary degeneration of the labyrinth.

Multiple sclerosis : (Ms)

Multiple sclerosis is a chronic, basically progressive disease of the central nervous system. Pathologically, a primary features of Ms is destruction of the Myelin sheath (demyelinization of nerve fibres sclerotic plaque formations may occur throughout the brain and spinal cord. The plaques are most numerous in the white matters of the cerebrum, brain stem, cerebellum and spinal cord. Clinically, Ms is characterized by episodes of localized central nervous system disorders cause of MS is unknown.

In individuals with MS, abnormalities of the auditory system may include disorders of the eighth nerve, brainstem, and temporal lobe. However, involvement of the cerebrum rarely occurs until a late stage of the disease (Merritt, 1963). Involvement of the eighth nerve is though to be limited to the CNS portion of the nerve.

Patient characteristics :

The initial onset of symptoms occurs between 20 and 40 years of age in approximately 65% of patients onset of symptoms below the age of 10 and above the age of 60 is uncommon.

MS is more prevalent in females than in males. The disease tends to become apparent at a slightly earlier age in females than in males. The female-to-male ratio is approximately 1.4:1.

Approximately 10% of patients with MS have another closely related member of the family affected with the disease. A hearing loss, if present, is bilateral in about 55% of individuals.

Site of disorder :

Eighth nerve and/or auditory pathways at the level of the brainstem. In patients with a relatively late stage of MS, a temporal lobe disorder may be apparent.

General Audiological Pattern :

Pure tone sensitivity results may vary widely inpatients with MS. As a broad rule of thumb, pure tone sensitivity results characteristically show a bilateral, SN hg loss for at least one freq. The degree of loss in generally mild. The audiometric contour frequently shows a sloping configuration with greater high frequency loss than low and mid frequency loss.

Impedance audiometry will show normal tympanograms, normal static compliance measures and abnormal acoustic reflexes. Reflex abnormalities may include reduced amplitudes, abnormal temporal patterns, or elevated or absent threshold measures, and abnormal acoustic reflexes. Reflex abnormalities may include reduced amplitudes, abnormal temporal patterns, or elevated or absent threshold measures.

Reflex threshold abnormalities may be characterised by a diagonal or horizontal pattern. In the diagonal configuration crossed and uncrossed acoustic reflexes are abnormal with sound to the affected ear. In the horizontal pattern. reflexes on both ears are abnormal in the crossed condition and normal in the uncrossed condition. The diagnoral pattern is consistent with eighth nerve site, the horizontal pattern is consistent with an intra axial brain stem disorder

Other abnormalities associated with MS may include deficits on masking level difference (MLD) tasks ; adaptation tests such as Bekesy audiometry. STAT and TDT ; and auditory brain stem evoked response (ABR) audiometry. In general, abnormality on auditory tests is more likely to be observed in patients with apparently wide spread central nervous system involvement.

Deafness in multiple sclerosis seems more likely to occur during the first 4 years of the presentation of the condition, but thereafter there is no relationship between the hearing loss and the duration of the disease. It has been estimated that some 3% of patients have a hearing problem but a higher percentage, perhaps 25% are troubled by vertigo at some stage during the disease. The disparity between the pure tone result which may be good and speech discrimination scores which are often poor is well recognized.

Because by definition multiple sclerosis is a disease characterized by multiple areas of demyclination of the central nervous system, the clinical diagnosis depends on the demonstration of two or more lesions. For this reason, non-invasive techniques of investigation are of particular value. Included among these are auditory and vestibular tests particularly brainstem auditory evoked, responses.

Cipparone et al. (1989) investigated the use of electronystagmography in 144 cases (116 definite and 28 possible) of multiple sclerosis. Pursuit movements and visual suppression tests were especially helpful being pathological in 56 and 58% of cases respectively ; spontaneous and/or evoked nystagmus Was present in 45%. Comparison between clinical and instrumental evidence of brainstem involvement indicated that 18% of the definite and 32% of possible cases of multiple sclerosis presented a negative clinical examination with positive instrumental findings.

Using a variety of tests, Jerger et al. (1986) examined 62 patients with definite multiple sclerosis. The acoustic reflex showed the highest identification rate (71%), followed by speech audiometry (55%), auditory brainstem responses or speech audiometry yielded a 90% identification rate. Auditory brain stem responses showed some abnormality in or both ears in only 52% of patients.

Wiegand and Poch (1988) investigated the acoustic reflex in normal controls. Patients with sensorineural hearing loss and asymptomatic multiple sclerosis. The threshold, onset latency and rise time measurements were similar for all three groups. Unlike normal subjects, there is no increase in wave V amplitude on binaural stimulation in a large majority of patients with multiple sclerosis who have no hearing deficit. The first stage of bilateral innervation occurs at the level of the superior olivary complex. Binaural stimulation may be complete at this level, so that only the subsequent waves originating from the brainstem nuclei caudal to the superior olivary complex will result in increased amplitude on binaural stimulation. Prasher. Sainz and Gibson (1982) showed that the mean amplitude of wave V in patients with multiple sclerosis

did not alter significantly when stimulation was changed from one ear to the other or even when both ears were stimulated simultaneously : the majority of their patients showed a decrease in amplitude on binaurai stimulation. In patients with chronic disease, the amplitude of wave V was small and was not affected by changing from monaural to binaurai stimulation. Their studies showed that the brain stem potentials in patients with multiple sclerosis who have no hearing deficit, did not increase in amplitude on binaurai stimulation. There is no single characteristic pattern either in this or other series but prolongation of wave V latency appears to be the most consistent finding.. By contrast Chiappa et al. (1980) reported brain stern auditory evoked responses in 202 patients with 'definite', 'probable', or 'possible' multiple sclerosis, but no patient presented with hearing difficulties. Only a few patients in their series had formal audiograms and all of these were normal. Using monaural stimulation, 68% had normal brain stem responses. In those showing abnormal responses, there was no significant correlation between the multiple sclerosis classification and the abnormality in brainstem response. In the abnormal group, 13% had only inter wave latency abnormalities. *55%* had only wave V amplitude, abnormalities, and 33% had abnormalities of both interwave latency and wave V amplitude.

Only relatively few patients are reported as developing acute hearing loss and two recent reports of such patients undergoing brain stem auditory evoked responses are of interest. Jabbari. Marsh and Gundersor (1982) reported two cases of acute unilateral deafness who responses showed an absence of waves II to IV in the first and the presence of only wave 1 in the second. Fischer et al. (1985) reported 12 patients with definite multiple[1] sclerosis who

experienced an acute hearing loss during a relapse of the demyelinating disease, in a series of 705 patients. Responses were recorded in all 12 patients, during the relapse with acute hearing loss in four and after the relapse with hearing loss in the remaining eight. During the relapse with hearing loss, brain stem electric response abnormalities were present in four wave I being absent in two. Responses were also noted to improve substantially when recorded after the relapse in two of the three patients in whom such records are made. Brain stem electric response recordings were abnormal on the side of the earlier hearing loss in five of the eigth patients investigated after relapse. Fisher et al. considered that the lesion causing unilateral hearing loss in multiple sclerosis could be situated in the cochlear nerve or close to its entry zone in the brain stem. In the classical case of a predominant, if not exclusive, central demyelination in multiple sclerosis, peak I remains present, well shaped and of normal latency in most patients who have not experienced an acute episode of hearing loss in the course of their disease. Fisher et al. reported an absence of wave I in only five cases in a series of 340 patients without a history of hearing loss. Arnold and Bender (1983) reported a case of particular interest in whom hearing tests were carried out over a 6 year period prior to the apparent development of multiple sclerosis. In spite of the subjective left-sided hearing loss, all the patients investigations including specialized tests showed no abnormality. Brain stem auditory evoked responses using monaural stimulation showed that the latencies beyond wave II were delayed, particularly wave V. One month after the investigations, the patient was struck and killed by lightning. Histopathological examination of the brain stem showed extensive demydination with specific sites of involvement in the superior olive, lateral lemniscus, and inferior colliculus.

Parving, Elberling and Smith (1987), using electrocochleography, showed prolonged action potential latencies and elevated thresholds at low intensity. At mid-intensities, there were significant deviations of action potential amplitudes. Hoff and Maurer (1983) examined a series of 71 consecutive patients suffering from clinically definite or clinically and probable multiple sclerosis. Wave I, as measured by early auditory evoked potentials, showed delayed latency with or without reduced amplitude in such patients and was interpreted as due to multiple sclerosis in eight. They concluded that the peripheral part of the acoustic nerve was involved in about 10% of multiple sclerosis patients.

Ferguson, Ramsden and Lythgoe (1985) sought to determine whether the combination of brain stem auditory evoked potentials and the blink reflex would yield a higher rate of abnormality than each test per formed separate!}. In a series of 50 patients with multiple sclerosis (definite - 30. probable - 10. possible - 10) using monaural stimulation, they found that 64% had abnormal responses. The blink reflex was elicited using electrical stimulation to the supra orbital nerve, fifty - two percent had an abnormal blink reflex, but when the results were combined with the brain stem electric response 76% were abnormal. In this series, symptomatic deafness was present in 20%.

Auditory temporal resolution has been examined in multiple sclerosis by Rappaport et al. (1994) and they gave a comprehensive review of literature. They found that multiple sclerosis patients exhibited a temporal processing defect and that their patients' performances suggested a predominant role of fore brain pathways in mediating auditory temporal resolution.

Mulsiek et al. (1989) investigated 33 subject with definite multiple sclerosis using seven behavioural and electrophysiological measures . 40% of the subjects with normal peripheral hearing complained of hearing difficulties.. They found the masking level diffference to be the most useful test diagnostically. It is known for its sensitivity for testing low brain stem / pontine lesions. They found the auditory brain stem response to be abnormal in 61.5% ; wave I was abnormal in five out of 26 (18%) ; wave III was absent in 25% of ears tested. Interwave latency measures I-III. III-V and l-V were not highly sensitive indices. Hendler, squires and Emmerich (1990) assessed central auditory function in 15 patients with multiple sclerosis. They confirmed the demyelinating lesions can cause a deficit in temporal processing. Abnormal masking level differences were always accompanied by abnormal auditory brain stem responses and mid-latency responses ; the subjects with abnormal masking level differences were more likely to have bilateral abnormalities in the auditory brain stem potentials. They also carried out MRI studies ; MRI signals restricted to levels caudal to the lateral lemniscus did not have abnormal masking level differences with the advent of MRI it has been possible to correlate electro physiological abnormalities including brain stem evoked responses and this has totally changed the diagnostic picture. Barratt, miller and Rudge (1988) published the report of one such patient who developed sequential hearing loss first on the right with recovery. MRi demonstrated lesions in the VIIIth nerve root entry zones which were thought to be responsible for the hearing loss ; they carried out two scans with gadolinium enhancement 1 month apart. Antoelli et al. 1988) examined 32 patients with definite multiple sclerosis, 21 (65.5%) showed auditory brain stem response abnormalities in whom 13 proved positive on MRI studies ; a

further two patients (15/32, 46.8%) also showed demyelination plaques in the brain stern. Other reports with MRI imaging include those of Cure et al. (1990) and Gstoettner et al. (1993).

Sudden deafness as a presenting symptom has now been recorded on a number of occasions and it is likely this number will increase as MRI becomes a routine procedure. Cases have been recorded by Shea and Brackmann (1987), Franklin, Coker and Jenkins (1989), Furman. Durrani and Hirsch (1989), Schweitzer and Shepard (1989) and Drulovic et al. (1993).

Multiple sclerosis has also been reported as the cause of sudden pontine 'deafness'. Drulovic et al. (1994) reported two cases presenting with unilateral sensorineural hearing loss and tinnitus. Brain stem evoked responses showed only the first three waves in the first patient and only wave I in the second. MRI showed foci of demyelination in the pons in case one and on the border between the pons and medulla in case two pure tone audionietry showed recovery after 1 month in both patients and remained stable during 1 year ; brain stem evoked responses however remained pathological after 1 month and again at 1 year Morgenstern and Kau (1988) had reported six patients demonstrating the clinical features of pontine deafness. The latter is characterized by an interruption of the central auditory pathway cranial to the olive. They emphasized that the pathogenesis of pontine deafness is still unknown but that it should be taken into consideration in the diagnosis of sudden unilateral deafness. In one patient they carried out positron emission tomography (PET). Tabira et al. (1981) reported cortical deafness in a patient with multiple sclerosis complete recovery from total deafness was seen

following stages of auditory agnosia and pure word deafness. ACT scan showed a transient low density area in the right parietotemporal region during the patients' second relapse. They pointed out that the most common cause of cortical deafness is cerebrovascular disease affecting both temporal lobes.

Mumps :

Mumps is an acute, contagious illness that usually causes (ever and painful non-purulent inflammation of the parotid glands. In many cases other organ systems involvement occurs such as meningo encephalitis, orchitis. panereatitis or deafness, The cause is usually the mumps virus (Paramyxovirus). however viruses such echovirus, coxsackie A and others have been implicated occasionally (Zollar and Mufsons, 1970).

There is an 18-21 day incubation period and a 1-3 day prodromal period of malaise, fever and discomfort over the angle at one or both jaws prior to the development of parotitis. During the next 24-28 hours one or both patotid glands becomes tender and swallen, often with surrounding jelly-like neck oedema. The ear lobe may be displaced laterally and the smooth line of the jaw is lost. The parotid duct orifice is inflamed, but if pus exudes from it the diagnosis of septic parotitis and not mumps should be made. The acute infection subsides in 72 hours though the salivary gland swelling may persist for 7-10 days. In some cases the parotitis may be absent. in others all four major salivary glands may become acutely enlarged and rarely only one sumandibular gland may be affected. This clinical picture has changed significantly since the introduction of the mumps vaccine. The peak incidence of mumps occurs in the 4-6 year old age group. The diagnosis is

made by demonstrating antibodies to the mumps S and V antigen and to the haemagglutination antigen. Studies have shown that more than *5%* of adults have neutralizing antibodies. The diagnosis may also be made by isolating the virus from urine, this can be performed upto 6 days before and 13 days after the salivary gland symptoms appear. Approximately 40% of attacks are

subclinical. Mumps meningitis is common but carries a good prognosis.
Cerebro Spinal Fluid (CSF) lymphocytosis occurs in approximately *65% of* affected patients but many do not show any symptoms. Management of mumps consists of good oral hygiene if the infection is associated with a history of fits, the temperature should be covered by tepid sponging and antipyretics such as paracetamol (but not aspirin).

Mumps is spread by aerosol droplets from the saliva and nasopharyngeal secretions of an infected individual and spreads easily in highly populated urban areas. In unvaccinated communities, epidemics occur approximately every 3-4 years. Apart from isolation of an index case, little can be done to prevent the spread of a mumps out break. Neither normal human immunoglobulin nor high titre mumps immunoglobulin is recommended. One attack confirms lifelong immunity. Mumps during pregnancy occasionally causes fetal endocardial fibroelastosis, or if contracted in the first trimester. abortion may occur. Termination of pregnancy is not usually recommended. Protective amounts of antibody across the placenta so that an, infant whose mother has already had mumps is unlikely to develop the disease in the first 6-9 months of life.

In the USA the incidence of mumps has fallen by 90% following the introduction of a live attenuated vaccine in 1967. In the UK theasles. mumps and rubella (MMR) vaccine has had an uptake of 85% since its introduction in 1985, resulting in the interruption of the 3 year epidemic cycle and has resulted in significant lowering of the incidence of mumps (Jones. White and Begg, 1991).

Neonatal Jaundice

Kernicterus (encephalopathy associated with severe unconjugated hyperbilirubinemia) has long been known to cause sensorineurai hearing loss. It is unclear whether the primary site of damage is peripheral, central, or both. In cochlear lesions, the audiogram usually reveals mild sensorineural hearing loss in the lower frequencies, gradually falling off to a severe hearing loss from 2000 Hz up. Rh or ABO blood-group incompatibility between mother and child is one of the most common causes, although several others. such as hepatic and biliary dysfunction, can be responsible. Acute bilirubin encephalopathy is most likely to cause damage between the third and seventh days of life, but damage may occur at older ages, even in adolescence. Regardless of the cause, hyperbilirubinemia should arouse suspicion of hearing loss. Hearing screening is recommended.

Renal failure :

The analogy between the nephron and the organ of corti is one which has become more frequent particularly with the increasing numbers of patients with renal failure who may be treated by haemodialysis or transplantation. It must be remembered that many of such patients have of necessity received

ototoxic drugs, either to control their infection or to promote diuresis. Certain studies therefore, are not only of interest but helpful to the otologist who is now more frequently involved with the management of such cases.

Yassin, Badry and Fatt-Hi (1970) found that the degree of hearing loss was directly related to the degree of hyponatraemia irrespective of the level of the blood urea. Urea by itself was non-toxic to the cochlear end organs and the effects on the cochlear were greatly improved by correcting the renal failure and restoring the serum sodium. Eighty percent of the cases with acute renal failure were improved by treatment, but only *52A%* of those with chromic forms.

Oda et al. 91974) have shown that in a study of 290 patients with chronic renal failure, 43 developed a significant hearing loss which could bo attributed to the therapy of the kidney problem. None of the patients were complaining of hearing impairment before the kidney treatment was started. Five patients treated with less than 60 haemodialysis showed no subjective hearing loss, three who had received more than 260 haemodialysis and multiple transplants complained of hearing and vestibular difficulties. During haemodialysis frequent and intense osmotic changes occur. Johnson and Mathog (1976) noted fluctuations in hearing in a single dialysis period, hut could find no correlation with corresponding changes in blood urea nitrogen, creatinine. Na, K, Ca, glucose, mean blood pressure level or weight.

Quiek (1976), in a prospective study of a large series of patients receiving dialysis and/or transplantation, found that a hearing loss occurs

quite frequently and while one factor might trigger off the loss, it was a combination effect of many factors, but this was not a simple addition of effects, more a potentiation. In his series, one in six had some form of hearing disorder six patients experienced sudden hearing loss and while a hypercoagulative state was evident in one patient, when the loss no apparent cause in the others.

Kligerman et al. (1981), in a prospective study of 67 patients with chronic end-stage r nal failure, noted a trend which appeared to suggest an association between haemodialysis and high frequency impairment, the degree of hearing loss did not vary with the length of treatment. Likewise, there was a striking similarity between the audiological findings obtained for all subjects with high frequency impairment, irrespective of medical treatment.

Hutchinson and Klodd (1982) assessed a series of 15 patients under the age of 60, Suffering from chronic renal failure who were being treated by has nodialysis.They eliminated from their study any patient who was diabetic of in whom the cause of renal failure was considered to be congenital. Each patient was tested once when the effects of the renal failure were most severe and they were about to undergo dialysis. They were tested using pure tone audiometry, acoustic reflex thresholds and reflex decay tests, Mectro nystagmography and brain item auditory evoked responses. They concluded that when ototoxic drugs, noise exposure, diabetis, congenital nephritis, and age above 60 years are eliminated, that although individual abnormalities will occur, chronic renal failure does not in itself produce a clinically significant hearing loss ; neither does it produce an abnormality of the peripheral or

central vestibular function that is clinically significant ; nor did it produce an abnormality with in the brain stem that affects the auditory or vestibular brainstem function from the clinical stand point.

A recent study by Gatland et al. (1991) investigated the prevalence of sensorineural hearing loss by pure tone audiometry in 66 patients with chronic renal failure and the threshold changes following haemodialysis in 31 patients. They found that the incidence of hearing loss was 41% in the low, 15% in the middle and 53% in the high ranges respectively. Of the 62 ears studied, 38% had a decrease in low frequency threshold after dialysis and 9% had an increase. They came to the-conclusion that fluctuation of the low frequencies with dialysis was common. Where a sensorineural hearing loss was found they carried out additional tests (loudness comfort levels and stapedial reflexes); these indicated the hearing loss to be of cochlear origin. Of their 66 patients. 51 were undergoing haemodialysis, seven continuous ambulant peritoneal dialysis, five were on diet alone and three under went renal transplantation. There was a history of gentamicin administration in 20% and metabolic bone disease existed in 66%. The effect of the high and low freq hearing losses produced a characteristic dome-shaped audiogram. Statistical analysis of the various parameters which they studied showed that age, derived plasma viscosity (from the globulin : albumin ratio) and gentamicin administration were significant factors in the actiology of the high frequency loss. However, none of the parameters measured was significant in relation to the low frequency loss they found no correlation between the fluctuation in hearing and weight charge with dialysis.

Kusakari et al. (1992) carried out a long-term follow-up study on the hearing of patients treated by haemodialysis. They investigated 37 patients with observation periods of 4 years or longer ; the average duration of the follow up study was 8 years 9 months (range 8 months - 11 months). Hearing tests were initially performed soon after the start of haemodialysis and every 3-12 months thereafter. Hearing corresponding to age was found in 43 ears whereas significant hearing loss was observed in 31 ears. The shape of the audiogram showed a high tone loss in the majority of cases and was bilateral and symmetrical in all except one case. They found two groups, the first in which the hearing deteriorated significantly is only three cases (live case) during the period of observation, but in these the hearing had completely recovered in two ears, partially in one with no recovery in the remaining two. In the second group the hearing was already reduced at the first test and remained unchanged in all ears except for one case in which both ears exhibited further deterioration. In two of their 37 cases with an initial hearing loss the cause was due to acoustic trauma in one and streptomycin toxicity in another.

Rh incompatibility

Human erythrocytes carries molecules ecoded by the rhesus (Rh) system. of which the RhD antigen is the strongest and the only one of clinical significance. There are two alleles at the RhD locus, D and d ; expression of D is dominant and about 85% of the British population are RhD. If a RhD woman becomes pregnant by a RhD father, the foetus may be RhD. At parturition, a significant leakage of fetal cells into the maternal circulation occurs which immunizes the mother to RhD. In the course of a second

pregnancy with a RhD fetus, a small number of fetal erythrocytes leak across the placenta and induce a secondary response in the mother. lgG antibodies to RhD cross the placenta, giving rise to haemolysis and jaundice. In severe cases, exchange blood transfusion with RhD blood is necessary' in ute.ro or following premature delivery.

To prevent sensitization of the mother, postpartum anti-RhD antibodies are given prophylatically. This hardramatically reduced the incidence of rhesus haemolytic disease in the new born, presumably because the anti-RhD antibodies remove fetal cells before they sensitize the mother. The decreased chance of rhesus haemolytic disease when the mother and fetus arc ABO incompatible can probably be explained in a similar fashion. The mechanisms involved in type II hyper sensitivity reactions may contribute to autoimmune disease in which organ-specific auto antibiotics are generated eg. and pernicious anaemia. It affects hearing, usually bilateral and most severe in the high frequencies.

Rubella :

Martin (1982) described rubella as being the commonest identifiable cause of congenital sensorineural hearing loss in children. Measles is the classic example of in utero disease producing severe sensorineural hearing loss. RNA virus which is present in the throat secretions, blood, and stool of infected persons. It probably enters the body by penetrating the upper respiratory mucosa. Congenital rubella is caused by transplacental transmission of the virus to the foetus. The disease is most common in children in 5-9 years of age, but many cases occur in younger children, adolescents, and young adults.

The incubation time between exposure and appearance of the rash of rubella is from 14 to 21 days. Head ache, fever malaise, lymphadenopathy and mild conjunctivities may precede the rash by as much as a week. particularly in adults. The exanthem often is the first sign of the disease in children. Rubella may cause lymph node enlargement alone, without skin lesions, and it may be unrecognized until serologic study results. Respiratory symptoms are not prominent. Forschheimer sports are small red lisions on the soft palate. These spots may be present but are not pathognomonic.

The rash of german measles is characterized by small, maculo popular. pink lesions, which are usually discrete. Sometimes they coalesce to form a diffuse erythematous exanthum. The rash starts on the forehead and face and spreads to the trunk and extremities. Usually, it is present for about 3 days and is preceded by tender lymphadenophathy which persists for several days after resolution of the rash. Post auricular and sub occipital nodes are most dramatically involved. Arthralgias and swelling of small joints may accompany the exanthematous period and may persist longer than other signs and symptoms. Purpura, hemorrhage, and encephalomyelitis also may occur.

Newton (1985) described a cohort of children in Manchester. She found that, although congenital rubella was relatively common, the number of children affected by it had declined compared to a previous study" of a similar population. The other significant finding in newton's series wa,s that 75% of the children with rubella deafness were born to mothers who had not been immunized. Davis (1993) also found that the part played by rubella as a cause of deafness in the UK had decreased. He noted however a ...large increase

in the number of pre term babies surviving with hearing impairment. Similar findings were reported by Parving and Hauch (1994) in a study of the causes of profound hearing impairment in a school for the deaf.

Rubella deafness is characterized by sensorineural hearing loss with a flat audiometric pattern. The severity of hearing loss may differ substantially of hearing loss may differ substantially between the two ears. Severe to profound deafness has been found in 4-8% of children with histories of maternal rubella, and it has also been recognized following asymptomatic maternal infections.

Deafness occurs in about one-third of rubella children. Affected children may also have microcephaly with mental retardation, eye lesions including cataracts and retinitis, abnormalities of the cardiovascular system and lower limb deformities.

There is a mistaken belief that deafness only occurs if infection is with in the first trimester. Hardy (1973) pointed out that infection with rubella at any stage in the pregnancy can cause deafness ; infection at 0-8 weeks - 53%, 21-35 weeks - 20%.

The virus enters the mother either through the nose or mouth and is transmitted through the placenta to the foetus. The maternal infection may be subclinical in about 40% of cases. Deafness is sometimes the only abnormality.

There is seasonal variation in the incidence of rubella (Martin. 1982). The numbers of children with rubella deafness born in December and January are much greater than those born in the summer months. This is not due to a seasonal variation in birth rate. It would appear that children conceived in March and April are more at risk of rubella than at any other time of the year.

Hemenway, Sando and McChesney (1969) described the abnormal findings in the ear. These may be abnormalities of the stapes or cartilaginous fixation of the stapes foot plate. The child's middle ear may contain fetal mesenchyme. The cochlea and saccule have Scheibe - type dysplasia.

The sensorineural hearing loss is usually severe to profound. Fisch (1981) described the typical loss as flat, affecting all frequencies more or less evenly, although it may be trough - shaped with a maximum loss for the middle frequencies. Wild et al. (1989) found the hearing loss to be flat with an average of 93dB. Only one child in their series of 57 had a progressive hearing loss.

Histopathology :

Rapid progress of the epidemiology and virology new knowledge of the histopathology of rubella deafness had remained fragmentary, because of the relatively small number of temporal bone specimens available [Friedmann, and Wright, 1966 ; Lindsay, 1973b] Microscopy of the cochlea showed Partial Collapse of Reissner's membrane with adhesence of the membrane to the stria vascularis and organ of corti. Small granulomas may be present between

the stria vascularis and Reissner's membrane. The tectorial membrane was found to be rolled up lying in the interval sulcus. Collapse of the saccule was observed, and the membrane was found to be collapsed and adherent to the macula saccule suggestive of a recent acute inflammatory process. There were only minor changes in the organ of corti. The hair cells were plentiful, as were the pillar cells, and appeared to be normal. There were some areas of cystic dilatation at the function of Ressiner's membrane and the spiral ligament.

The granulornatous lesions described by several authors appear to have occurred when the organ of corti had reached morphological maturation [Friedmann and Wright, 1966 ; Bordley and Hardy, 1969 ; Brook hauser and Bordley, 1973 ; Lindsay, 1973a). This could be interpreted as consistent with the degeneration of the performed neuroepithelial structures, reflecting continued vinus cell interaction, as suggested by other, stigmata of the rubella syndrome. Unusually large granulomas have been noted in the stria vascularis of the cochlea from an 8 year old deaf boy. His mother had suffered from an unrecognized, viral infection, probably rubella in the fourth month of her pregnancy. The boy suffered from deafness and renal disease. At postmortem (Dr. A.C. Cameron, consultant pathologist, the Children's Hospital. Birmingham), various features of the rubella syndrome were identified : there was absence of the falx cerebri and the ductus arteriosus was patent. The serum antibody titre for rubella was higher than 1:512.

Prenatal rubella is recognized as a cause of congenital deafness, but its importance may not be fully appreciated (Brook hauser and Bordley. 1973).

One reason is that a woman may have a silent rubella infection during preganancy and pass the virus to the foetus without any clinical evidence of her own infection (Alford, Neva and Weller, 1964 ; Menser, Dods and Harley. 1967). Of 84 pregnant women infected with the rubella virus, but without clinical disease, 10 gave birth to children from whom the virus was isolated (Bordley et al., 1968). A study of the effect of rubella on the frequency of congenital deafness for 5 years after an epidemic in 1960, in an island population, revealed that of 87 congenitally deaf children born the year after the epidemic, all but one had suffered deafness as the only demonstratable congenital abnormality. Only 20 gave a history of first trimester rubella, so by the usual classification the remaining 67 cases would be labelled as idopathic, all known causes having been ruled out. However, serological tests for rubella antibodies on 30 of the 'idiopathic' deaf children were positive in 74% compared with 30% in a control group born with in the same year (Karmody, 1968).

In the investigation of congenital deafness in a child, a test for rubella antibodies should be performed. With increasing age a positive result become less significant, but the absence of rubella antibodies should be performed. With increasing age a positive result becomes less significant, but the absence of rubella antibodies would exclude the virus as a cause and focus attention on other as a cause and focus attention on other factors.

The affinity of certain viruses, in particular, the paramyxoviruses and tocoviruses, for the cochleo vestibular system has been recognized. There is mounting evidence of a viral infection in otosclerosis. The application of

immuno histochemical methods has revealed the expression of antigens of rubella, mumps and measles in the otosclerotic foot plated examined (Arnold and Friedmann, 1988). The viral antigens are more strongly expressed by the cells of the perivascular tissue in oto spongiosis and by various inflammatory cells and also osteoclasts in the resorption lacunes of oto sclerosis (Friedmann and Arnold, 1993).

Diagnosis :

The diagnosis of rubella is often made on clinical grounds. It is possible to culture the virus from throat swabs or samples of stool or urine upto the age of 6 months. Persistence of IgG antibody after the disappearance of maternal IgG indicates congenital infection. Rubella specific IgM is present in the infected child for about the first 6 months after birth. Newton (1985) pointed out that the earlier the hearing loss is diagnosed and investigations commenced, the greater the likelihood of concluding that the hearing loss is due to a congenital infection such as rubella.

Martin (1982) pointed out that eradication of rubella would abolish one-fifth of all congenital sensorineural deafness. The policy in the UK at present is to offer vaccination to all girls between 10 and 14 years of age and also to screen all women at antenatal clinics. There are several problems with this policy. For it to be successful in abolishing congenital rubella there would have to be almost a 100% uptake in the target population. This is known not to be the case. In addition, the vaccine would need to be 100% effective (Begg and Noah, 1985). At present no check is made on girls after vaccination. Around 40% of babies damaged by rubella are first born. Antenatal

screening for rubella is therefore too late since the foetus may already be infected (Kudesia et al., 1985). These authors advocated a change a policy with testing before and after vaccination to ensure a primary response. This technique would also distinguish women who were protected by the vaccine from those with antibodies to the natural virus.

Rubeola and Other Infections

Measles (rubeola), cytomegalic inclusion, disease, herpes, roseola. infectious mononucleosis, varicella, Mycoplasma pneumonia, typhoid fever, scarlet fever, influenza, and other infections have also been associated with sensorineural hearing losses. The hearing loss may be severe or profound and may be sudden or gradually progressive. So far, only symptomatic and preventive therapy is available. These disease may occur in adults, in children and in utero. Particular effort should be made to protect pregnant mothers from exposure to these infectious agents Measles and scarlet fever also are notorious for their destruction of the eardrum and middle ear.

Scarcoidosis :

Scarcoidosis is a rare systemic granulomatous disease of unknown aetiology. Head and neck manifestations are uncommon and when encoutered in otolaryngological practice, the disease usually involves the parotid gland. facial nerve, nasal cavity and larynx. The nervous system is affected in only 5% of cases, although this rises to 50% if uveoparotid fever is present. The central nervous system lesion is presumed to be a granulomatous meningitis that directly infiltrates the cranial nerves or causes them to be compressed from involvement of adjacent intracranial structures. Any of the cranial nerves

may be affected but the facial nerve is most frequently involved while the VIIIth cranial nerve is fourth in order (Hybels and Rice, 1976). The disease has a higher prevalence among Blacks in America.

The organs most frequently affected are the lymph nodes, lung, liver, spleen, skin and eyes, but tissue may be involved and certain manifestations are known to be associated with particular HLA types. The course of the disease is usually chronic with minimal constitutional upset.

Serum angiotensin - converting enzyme (ACE) levels are raised in nearly two-thirds of cases of active sarcoidosis, but false positive elevation of this enzyme can occur. False positives, however, are extremely rare in the Kveim test. De Remee and Rohrbach (1980) noted that serum angiotensin - converting enzyme levels closely paralleled and occasionally antedated changes in clinical status in patients either undergoing spontaneous remission or being treated with steroids and suggested that enzyme determination should be of value in management. However, serum angiotensin - converting enzyme levels may also be raised in other conditions, such as Gaucher's disease and leprosy.

All patients with the disease should have assessment of their liver and renal function. The alkaline phosphatase is frequently raised and may be due to involvement of either liver or bone. Approximately 5-10% of patients with sarcoidosis have elevation of their serum calcium and this is thought to be due to hypersensitivity to vitamin D. There is hypoglobulinaemia in about 25% and this may also reflect disease activity. Electrophoresis of the serum proteins usually shows increased α-2 and *y* globulins. The full blood count

is frequently normal but the erythrocyte sedimentation rate may be raised in the active stages. It is a characteristic feature of sarcoidosis that infiltration of old scars often occurs and these may provide welcome biopsy material.

Otological features :

Sarcoidosis involving the ear may be associated with other signs such as uveitis (80%), parotid swelling (20%), facial nerve palsy (439;) and lymphadenopathy (55%). However, 40% of cases have shown no other neurological involvement, and isolated VIIIth nerve disease has also been reported (Souliere et al., 1991).

The hearing loss may be sudden, fluctuating or progressive and the degree may vary from slight to severe to even total loss. It is usually bilateral although one side is frequently may show either a high or low frequency loss while caloric testing usually shows reduced or absent responses (Gristwood. 1958 ; Hooper and Holden, 1970 ; Kane, 1976). The pathogenesis of the hearing impairment is undecided. From the 50 or 80 recorded cases it would appear that the hearing loss is most probably sensorineural, but electro cochleography in two cases suggested the lesion may be retrocochlear with normal hair cell function (Majumdar and Crowther, 1983). One of the cases reported by Souliere et al. (1991) had a cerebellopontine angle granuloma that mimicked a vestibular schwannoma.

The temporal bones from a 32-year-old man, deaf for 5 years from central nervous system sarcoidosis, have been examined histologically (Babin. Liu and Asbenbrener, 1984). It was found that the acoustic, vestibular and

facial nerves were involved in a striking perivascular lymphoeytic infiltration resulting in myelin and axonal degeneration. The cochlear and labyrinthine neuroepithelium and stria vascularis had degenerated. Babin, Liu and Ashenbrener hypothesized that sensorineural deafness and vestibular dysfunction in sarcoidosis start as a reversible neuropathy ; in some patients an ischaemia secondary to the vasculitis results in irreversible damage to the inner neuroepithelium.

Steroids remain the mainstay of treatment but their effectiveness is not assured especially in those with a profound or total hearing loss. More recently immunosuppressive drugs have been used though because of the few cases treated their efficacy is as yet unproven.

Sickle Cell Disease

In the United States, about 7-9% of blacks carry sickle cell traits. About 1 in 400 has sickle cell disease, an autosomal recessive condition. Anemia, splenomegaly, and attacks of abdominal pain, jaundice, weakness. and anorexia develop. Sensorineural hearing loss occurs in about 20-25% of patients with the disease. Pathology in the inner ear is consistent with ischemic changes and is believed to be due to thromboembolic disease secondary to sickling. Sudden total deafness also has been reported and is believed to be caused by a vascular occlusion. In some cases, serve sensorineural hearing loss associated with sickle cell crisis has spontaneously returned to normalcy. Treatment is generally symptomatic.

Stroke

Hemorrhage into the ear produces deafness, as noted in the discussion of coagulopathies. Similar findings occur following spontaneous subarachnoid hemorrhage, which produces blood in the internal auditory canal and cochlea. Major cerebrovascular occlusions may produce severe hearing deficits. Occlusion of the vertebral of posterior-inferior cerebellar artery produces lateral medullary syndrome, or Wallenberg's syndrome. This syndome is characterized by ipsilateral ptosis and miosis; enophthalmos; facial hypesthesia: palatal. pharyngeal, and laryngeal paralysis; contralateral hypesthesia and decreased thermal sensation in the trunk and extremities; as well as occasional involvement of the sixth, seventh, an eighth cranial nerves. Sensorineural hearing loss occurs, and vestibular function is abnormal.

Occlusion of the anterior vestibular artery alone produces vestibular symptoms without hearing loss. Occlusion of the anterior-inferior cerebellar artery generally produces sudden vertigo with nausea and vomiting, hearing loss, facial paralysis, and cerebellar and sensory disturbances. Degeneration of the membranous labyrinth and brain stem auditory and vestibular nuclei occurs. Ipsilateral loss of pain and temperature sensation on the face are common, associated with decreased pain and temperature sensation on the opposite side of the body. Patients who survive usually improve slowly.

Vertebrobasilar ischemia may have similar, but transient symptoms, of which vertigo is the most prominent. Other associated findings may be hearing IQSS, diplopia, headaches and speech difficulties. Although atherosclerotic vascular disease is the usual etiology, arthritis, syphilis, aneurysms. and subclavian steal syndrome also must be kept in mind.

Lateral venous sinus thrombosis and thrombophlebitis of the jugular bulb or the internal jugular vein may cause mastoid infection, brain abscess, or septicemia and meningitis due to spetic emboli, and they may lead to hearing loss. In the past, these diseases generally have been seen as complications of ear surgery. Recently, however, jugular thrombophlebitis and its complications (including retrograde extensions) have been seen in heroin addicts who use the subclavian or internal jugular veins as access routes.

Syphilis

The effects of syphilis on the temporal bone are now seen very much less frequently in clinical practice. It is a disease which should, nonetheless, be suspected in any patient presenting with tinnitus and / or vertigo and / or sensorineural hearing impairment, particularly if fluctuant & of sudden onset prompt recognition & treatment may halt or possibly reverse the progressive audiovestibular symptoms, & prevent the development of serious systemic involvement in the territory stage if this is not already present. These serious systemic features include cardiac & aortic involvement & parenchy-matous neurosyphilis, manifested by general paralysis of the insane & tabes dorsalis. Both the congenital & acquired forms of syphilis can be complicated by inner ear disease.

The last half century has witnessed a dramatic decline in the number of reported new cases. Thus the incidence of new cases of congenital disease in the UK fell from 2439 in 1931 to 1223 cases in 1950 and 150 in 1974. In .1980, only eight new cases were diagnosed in children under 2 years of age (British medical journal, 1982). The universal antenatal serological

screening programme in the UK has undoubtedly played an important part in thecontrol of congenital syphilis ; an untreated syphilitic mother has about a 50% chance of beanng a syphilitic child. Failure to eliminate this from, of the disease altogether is probably due to the difficulty in administering antenatal care to some social groups. During the same period, the overall reported incidence of new cases of both congenital and acquired types fell from the post-war peak of nearly 28000 cases per year to about 4500 cases in 1980. The prevalence in 1984 was 6.4 cases per 100000 (British Medical Journal. 1986), but is now even lower due to the impact of AIDS on contemporary sexual practices. Currently well over 50% of syphilitic infections in men are reported to have been homosexually acquired. The male : female incidence is now about 4:1 and new cases of acquired syphilis are about, 25 times more frequent than congenital ones.

Diagnosis :

Of the established screening tests for syphilis, the venereal disease. Research Laboratory (VDRL) slide test is probably still the one most commonly undertaken in clinical practice. Although the test is frequently negative in previously treated cases, & false positives may occur, it does give an indication of disease activity. It is invariably strongly positive in high dilution in early untreated cases and is usually accompanied by an elevated crythrocyte sedimentation rate.

More specific serological tests are now routinely employed, e.g. Treponema pallidum haemagglutination test (TPHA) ; Treponema pallidum immobilization test (TPI) and the fluorescent treponemal antibody absorption

test (FTA-ABS). Of these the FTA-ABS is the most sensitive (Hughes and Rutherford, 1986). A positive result confirms previous syphilitic infection but does not reflect disease activity and stays positive even following adequate treatment. It is currently common practice to diagnose otosyphilis in any patient with inner ear of unknown cause and a positive FTA-ABS test result However, a recent survey by Hoare et al. (1996) reported the results of a prospective study of syphilis serology in nearly 1800 new otolaryngological out patients ; 40 (2.2%) were positive. None had congenital or neurosyphilis, and as many had non-otological symptoms unrelated to their serological status as had cochleovestibular symptoms. Hughes and Rutherford (1986) have also highlighted the clinical dilemma caused by limitations in the predictive value of the serological tests for syphilis. A solution has recently been proposed by Birdsall, Baughn and Jenkins (1990), who advocated a new western blot assay to eliminate the possibility of a false positive result and to confirm whether the disease is active.

Examination of the cerebrospinal fluid in patients with syphilitic ear disease is desirable to look for possible evidence of central nervous system involvement which is more likely to be seen in the late acquired form. Typical cerebrospinal fluid abnormalities of neurosyphilis, apart from positive serological test, include slightly raised globulin and IgG levels and a lymphocylosis. Such investigations and treatment are best coordinated by a venereologist, who will also need to examine possible contac's in cases of acquired syphilis. Gleich, Linstrom and Kimrnelman (1992) found that the otovestibular symptoms of patients with syphilis and cerebrospinal fluid abnormalities invariably improved with high dose penicillin and steroid treatment.

General features :

Congenital syphilis may be associated with other abnormalities outside the cochleovestibular system. The ocular manifestations of interstitial keratins and choroidoretinitis result in corneal capacity is about 90% of patients with otological symptoms. Such features may only be apparent on careful slit - lamp examination by an ophthalmologist but can be of diagnostic value. Hutchinsonian thickened wedge-shaped incisors which are occasionally notched are found in 20% of cases. The typical facies of frontal bossing of the skull due to involvement and collapse of the nasal septal cartilage and bone arc only present in about 10% of cases (Morrison, 1975 ; Belal and Linthicum, 1980). Other features such as 'Sabretibia' are rare.

Tabes dorsalis and general paralysis of the insaneare manifestations of neurosyphilis and both are now rare. The neurological features include 'lightening' pains, early optic atrophy, Argyll Robertson pupils, bladder dysfunction and sensory loss from dorsal column involvement resulting in impaired vibration sense and joint disruption - charcot's joints (Catterall, 1977). However, previous treatment which may have been inadequate often results in a typical features.

A not infrequent clinical dilemma is posed by patients from the West Indies, Central America and Africa, who may display positive serological test results and similar clinical manifestations but who are suffering from yaws. This disorder is caused by a different spirochaete. Treponema, pertenue. and typically is spread by direct contact among children. Old scarring from previously healed cutaneous ulcers is characteristically present on the lower

legs. When these scars are absent, a patient from these countries should certainly be considered to be suffering from syphilis and treated accordingly.

Gtological features :

Histopathology :

Two distinct types of histopathology are recognized. Treponemal labyrinthitis is the typical lesion in early congenital syphilis, and meningolabyrinthitis in the acute meningovascular phase of secondary and tertiary disease. In this latter form, the small blood vessels of the meninges show endarteritis obliterans. There is increased fibrosis of the meninges. with small areas of necrosis and a diffuse infiltration by plasma cells and lymphocytes. The VIIIth nerve may be involved in associated with the infective basal meningitis, and the inflammatory process spreads from the spiral ganglion to the cochlear duct and membranous labyrinth (Goodhill, 1939).

It late congenital and acquired disease, the main lesion is, a rarefying gummatous osteitis of the temporal bone with secondary involvement of the membranous labyrinth (Mayer and Fraser, 1936 ; Goodhill, 1939: Schuknecht. 1974). All three layers of otic capsule are involved in the osteitis, which is associated with underlying endarteritis and infiltration with chronic inflammatory cells and multinucleated giant cells. The inner ear features are dominated by endolymphatic hydrops and progressive degeneration of the neuroepithelial structures, particularly the cochlear neurons and organ of corti. which may be severe.

It has long been held that the pathogenesis of the hydrops is probably by direct involvement of the endolymphatic duct which becomes obliterated. However, treponemal spirochaetes have been found in many different sites in humans with late syphilis following treatment, including aqueous humour of the eye, cerebrospinal fluid, synovial fluid, temporal artery, lymph nodes and liver (Smith and Israel, 1967 ; Mack et al., 1969). This continued presence of spirochaetes, in spite of apparently adequate previous antibiotic treatment, may well be a significant factor in the pathogenesis of the hearing loss.

Early syphilis :

Congenital syphilis is contracted by the developing foetus in utero as a consequence of acquired maternal syphilis. The early infantile form is usually fatal due to multisystem involvement which dominates the features of otolabyrinthitis. As noted above, it is now exceedingly rare in the UK. Probably about 50% of cases develop bilateral hearing loss eventually. Earlier studies, e.g. Karmody and Schuknecht (1966) tended to under estimate the incidence because of the proportion of younger individuals who could be expected to develop symptoms later on.

Secondary syphilis is typically, although not exclusively, seen in adult homosexual men. The first symptoms last for a few weeks and include malaise. slight pyrexia, non-specific headaches, skin eruptions, pharyngitis and lymphadenopathy. They are relatively trivial and are hence frequently ignored by the patient until sudden hearing loss develops which is often bilateral. There may be some transient vestibular symptoms, which are frequently positional in character, and tinnitus. Ocular palsies and facial paralysis may

occur as well in the acute maningo vascular type of secondary disease. The sensorineural hearing loss preferentially affects the high frequencies : elevated stapedius reflex thresholds, possibly with reflex decay, are frequently present speech discrimination is often significantly worse than is suggested by pure-tone audiometry and the caloric responses are reduced. Increased latency and/ or reduced wave V amplitude on brain stem evoked audiometry has been reported (Rosenhall, Lowhagen and Roupe. 1984). These audiovestibular symptoms may be partly reversible. It left untreated, the infection tends to run a benign course but the hearing loss remains.

Late syphilis :

Late syphilis affects the temporal bone between 10 and 50 years after the primary infection. Once established, the untreated disease carries a poor prognosis with relentless progression to profound deafness, although fluctuations are common. There are some grounds, however, for optimism with antitreponemal agents and systemic steroids.

In general, the clinical features are similar in both the congenital and acquired forms of the disease, although the former is more common in women. It is often difficult to assign a patient to one of these groups, particularly since previous antibiotics have invariably been taken. The otological features in congenital cases can occur at any stage, but they are uncommon after middle age. In contrast, patients with late acquired disease are usually over 40 years of age. The hearing loss is typically symmetrical in congenital cases but more frequently unilateral in the acquired group, sometimes for many years. In about 20% the onset of aural symptoms is sudden and

fluctuations are seen in 30%, particularly in the early stages (Hahn. Rosin and Haskins, 1962 ; Dawkins, Sharp and Morrison. 1968 : Kerr. Smyth and Cinnamond, 1973). Apart from the. fluctuation, there are other features which closely mirror the symptoms of Meniere's disease and are a reflection of the underlying endolymphatic hydrops (Schuknecht. 1974). The early hearing loss is sensory in character with predominantly low peaked patterns of pure tone audiometry. Half the patients exhibit episodic attacks of vertigo which may be indistinguishable from those occurring in classical meniere's disease.

The results of transtympanic electrocochleography in a series of 18 cases of late syphilitic deafness have been described by Ramsden. Moffat and Vibson (1977). An enhanced negative summating potential was found in nearly 80% of ears tested in association with a small cochlear microphonic. both features indicating established endolymphatic hydrops. The summating potential characteristically affected the descending link of the compound action potential. This feature, however, is not pathognomonic and is the authors experience occurs relatively infrequently. Nagasaki et al. (1993) also found evidence of established hydrops on electro cochleography in a large proportion (56%) of patients with syphilitic labyrinthitis. Syphilitic hydrops tends to remain relentlessly active in the majority of cases, in contrast to idiopathic Meniere's disease where only a relatively small proportion of patients have hydrops symptoms which are not self-limiting to some degree. Secondary neuronal degeneration associated with more profound degrees of hearing loss is therefore more frequent. The pattern of pure tone audiometry now becomes flattened or high tone in character. Alteration of the stapedius reflex to a retro cochlear

pattern with elevated thresholds and decay is now evident in associated with a relative greater impairment of speech discrimination.

Progressively severe peripheral vestibular damage leading to increasing in balance and ataxia is also quite common. However, compensation for such a slowly developing deficit can significantly reduce the degree of disability, particularly in the younger patient, and may only come light on formal vestibular assessment.

Two ponymous otological phenomena which are sometimes present in late congenital syphilitics are worthy of mention. Hennerbert's (1911) sign consists of a transient positive fistula test without clinical evidence of middle ear disease. Tullio's sign consists of transient vertigo and nystagmus following exposure to sudden high intensity sound. These phenomena are believed to be due to sound energy transmission through the stapes footplate on to the distended saccule, and are occasionally seen in other diseases associated with endolymphatic hydrops.

Treatment :

Penicillin is still the most effective antibiotic for the treatment of syphilis. Its main bactericidal effect occurs when the organisms are dividing this has been shown to take place much less rapidly in the late form of the disease, and hence the duration of treatment is as important as the maintenance of effective serum concentrations. In the presence of confirmed allergy, one of the cephalosporins is probably the second drug of choice.

The proven effective therapeutic regimen consists of 600000 units of procaine penicillin by intramuscular injection daily for 21 days. This aqueous solution only has to be injected once a day and results in an effective serum level for 24 hours (Catterall, 1977). Oral probenecid 500mg 6 hourly inhibits exerction of the drug and helps to raise tissue levels. This regimen has proved satisfactory for out patient treatment (Dunlop, Al-Egaily and Houang. 1981). An alternative protocol which is probably as effective in patients who show good treatment compliance is high dose ampicillin. A dosage of 1.5g is prescribed four times daily for 4 weeks (Adams et al., 1983). Unfortunately, there is no evidence that penicillin treatment alone prevents the progression of cochleo vestibular manifestations.

There is now, however, considerable clinical evidence that systemic steroids alone can improve the hearing at least temporarily, in upto 50% of cases with late syphilitic deafness (Hahn, Rosin and Haskins, 1962 ; Karmody and Schuknecht, 1966 ;iMorrison, 1969 ; Kerr, Smyth and Cinriamond. 1973) and suggests an immunological basis for at least part of the hearing loss. Steroids are also indicated to prevent the adverse effects of a possible Herxheimerreaction. This is a systemic phenomenon occurring within 2-12 hours of the first antitemponemal injection and is characterized by fever, followed by headache Malaise, flushing and Sweating. The reaction lasts for a few hours and is often accompanied by worsening local tissue involvement and has been known to cause sudden increased hearing impairment. The reaction has been attributed to complement activation and to complex immunological reactions involving a hypersensitivity response to the

disintegration products resulting from sudden destruction of large numbers of spirochaetes (Catterall, 1977).

Prednisolone 30mg 8 hourly is therefore commenced prior to institution of antitreponemal treatment and continued for 4 weeks. Others have preferred to use ACTH (Kerr, Smyth and Urinamond, 1973 ; Adams et a!., 1983). If there is no evidence of improvement in the auditory and vestibular symptoms by 6 weeks, it is discontinued. Improved hearing thresholds are more likely in patients with fluctuant symptoms and are an indication for, longer term treatment on a maintenance dose of 2.5-5mg daily. Unfortunately, any hearing gains often relapse on withdrawal of steroids which may well therefore need to be taken on a long-term basis to maintain improvement. Of course, prlong steroid treatment has well recognised side effects and decisions to maintain steroids must be weighted carefully in each individual case. Discontinuation of steroids should be followed by a further course of antibiotics. Initial optimism about the successful out come of treatment of late syphilis with penicillin and steroids has been tempered in recent years, although long-term results show that this regimen frequently prevents further hearing impairment and almost invariably preserves some hearing (Adams et al.. 1983 ; Chan. Adams, and Kerr, 1995).

Treacher Collins and Franceschetti-Klein Syndromes

Mandibulofacial dysostosis was first described in the 1840s. and the facial appearance is classic hypoplastic zygomas produce downward-sloping palpebral fissures. Cheekbones are depressed, the chin recedes, the mouth has a large "fishlike" appearance, the mandible is hypoplastic, and coloboma of

the lower eyelids with lack of cilia is common. Auricular malformations occur in approximately 85% of such patients.About one-third have external auditory canal atresia or an ossicular defect. Conductive hearing loss is most common, although sensorineural deafness has also been reported. Surgical treatment is rewarding in carefully selected patients, but early amplification should be use in patients with bilateral hearing loss. The inheritance pattern is autosomal dominant.

Tuberculosis

Increasing numbers of patients with tuberculosis are currently presenting of various specialist departments in the UK, most often among immigrant communities, unfortunately it can no longer be considered a disease of the past. Although the infection primarily affect the middle ear, it may cause secondary involvement of the bony labyrinth.

Otological features

The possibilities of tuberculous involvement is usually entertained by the presence of certain a typical features of chronic suppurative middle ear disease windle - Taylor and Bailey (1980) comprehensively reviewed a series of 22 patients with tuberculous ear disease who presented to the Royal National Throat, Nose and Ear Hospital, over a 30 year Period and found one half to be under 20 years of age. None had a past history of pulmonary tuberculosis, although 18% had previously diagnosed disease at other sites. The middle ear features are dominated by the presence of florid, pale granulation tissue, which is often visible as a 'mass' behind the tympanic membrane. Occasionally, as in other granulomatous disorder the tempanic membrane may be intact, but

more often breakdown has occurred, characteristically resulting in multiple perforations. Co-existent secondary infection by other organisms is frequently found. Yaniv, trailb and conradie (1986) subsequently reviewed a series of 24 cases of otological tuberculosis and reported similar findings.

Concomitant sensorineural hearing loss is encountered much more frequently than in 'conventional' Chronic suppurative otitis media, and often results in a disporpostionately large hearing loss. Windle-Taylor and Bialey (1980) did not detail the precise Sensorineural hearing loss, but their data indicated that 60% had inner ear involvement, and in 25% this loss was total.

Treatment

Management obviously involves surgical excision ad drainage of middle ear and mastoid disease in conjunction with antituberculous treatment. As in patients with syphilis referral to a physician for general assessment co-ordination of medical treatment and tracing of possible infective contacts is mandatory. Although these have been isolated reports of ototoxicity by rifampicin and ethambutol the risk is very considerably lower than following steptomycin therapy which has therefore been largerly discontinued. After the infection has been controlled by chemotherapy, any residual tympanic membrane defects can be managed successfully by conventional tympanoplastic surgical. techniques (Ma. Tang & Chan, 1990).

Vasculitis

Rhenumatoid arthritis, giant-cell arteritis, polyarteritis nodosa, leukocytoclastic angitis, and various other vaculitides have been associated

with hearing loss. Middle-ear fluid with conductive hearing loss is common, and sensorineural hearing loss also may occur. Occasionally, middle-ear disease may precede other manifestations or a vasculitis syndrome, or it may persist following otherwise successful therapy with steroids or other medications. In patients with known systemic vasculitis and conductive hearing loss that does not respond to conventional therapy, exploratory tympanotomy and middle-ear biopsy may be indicated. This combination may be performed as a diagnostic measure early in the course of the disease if primary vasculitis is suspected. Early detection and prompt treatment are the mainstays of therapy.

Vascular Disease

Patients with advanced atherosclerosis, particularly those who have suffered myocardial infarctions, have a higher incidence than the normal population of high-frequency sensorineural hearing loss. The pathogenesis is undetermined, but it is believed to be related to vascular changes within the inner ear. The subclavian steal syndrome involves collateral circulation from the vertebral artery in the presence of proximal left subclavian artery stenosis. Hearing loss occurs in nearly 10% of patients and results from compromise of the vertebrobasilar system, which provides blood to the inner ear. Other otologic symptoms, such as vertigo and facial paralysis, also may occur with this syndrome because the vertebral artery supplies the pons, medulla, cerebellum, vestibular and cochlear labyrinth, and portions of the temporal lobe, as well as the upper spinal cord, thalamus, and occipital cortex. Similar, symptoms may occur with more limited dysfunction of the vertebral system, such as the lateral medullary infarction syndrome. Surgical treatment is available.

Sudden hearing loss is often ascribed to "vascular causes". Although this explanation is tempting, histological confirmation s scarce, although the phenomenon certainly exists- at least in association with larger cerebrovascular occlusive events. Many disease which may cause anoxia of tissue may be responsible for sensorineural hearing loss, but more research is needed to prove the relationship. Such conditions include chronic hypotension, anemia. vasoyagal abnormalities, and other similar maladies.

Waardenburg's Syndrome

This dominant syndrome includes partial albinism (classically seen as a white forelock of hair), laterally positioned medial canthi, different colored irises, and congenital non-progressive sensorineural hearing loss. Vestibuiar abnormalities and temporal bone radiological abnormalities may occur. The deafness may be total with only slight residual hearing in the low frequencies: moderate, with near-normal hearing in the higher frequencies and severe loss in the low frequencies; or unilateral with near-normal hearing on one side. Only 20% of patients with Waardenburg's syndrome demonstrate hearing loss; however, Waardenburg's syndrome accounts for about 1% of all hereditary deafness. At present, no treatment exists other than sound amplification when applicable. Genetic counseling is relevant in these cases.

SUMMARY AND CONCLUSION

Hearing loss may accompany many systemic diseases. Familiarity with the otologic manifestations of these conditions facilitates early diagnosis and treatment of hearing impairment. Moreover, attention to these relationships often leads to the diagnosis of other wise unsuspected, potentially serious systemic diseases in patients who complain of hearing loss.

Fortunately, many of the hereditary causes of hearing loss are preventable or treatable. Most of the syndromes involve known inheritance patterns, which makes genetic counseling useful in their management. Recessive syndromes can be minimized by avoiding consanguineous marriages screening programms. Such as for sickle cell trait and Tay - sachs disease, are also helpful.

Many of the in born errors of metabolism and chromosome anomalies can be detected in utero by amminocentesis, allowing for possible elective abortion. The effects of maternal and paternal advanced age and other factors associated with increased appearance of congenital anomalies can be minimized as physician and patient populations become more familiar with these disease and their become more familiar with these disease and their causes.

The great many diseases that may be associated with hearing loss highlight the need for a through history and physical examination in each patient with hearing impairment, and to concentrate more on the prevention of disorder in infants as a audiologist & speech & longnage pathologists.

Moreover, they remind us to search for unsuspected hearing loss rarely in patients with these maladies. Much more information is needed to classify the nature of hearing loss associated with systemic diseases. Of particular importance is the need for temporal bone specimens for further research only through constant clinical attention and diligent investigation can we hope to diagnose, understand, and prevent hearing loss of all causes.

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