PERCEPTIVE BASS DEAFNESS

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PROEFSCHRIFT

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INTRODUCTION

In 1948 FOWLER still wrote in his manual "deafness from neural lesions is never limited to the low tones" and "only if there is an accompanying conduction lesion, is it possible for the slope of the hearing curve to be upward from left to right".

Perceptive bass deafness does occur so often, however, that it seems that FOWLER's pronouncement, even in its generality, cannot be accepted at all.

When an audiogram is made of a patient who complains of deafness, and the hearing loss proves to be greater for the low frequencies than for the high ones, it may be said that in such a case the deafness is characterized by a rising audiogram. Yet in such cases we cannot simply speak of a bass deafness. In the audiogram a real bass deafness shows a hearing loss for the low frequencies, while the hearing for the high frequencies is normal or practically normal.

There are some exceptions to this rule. A bass deafness may occur in an auditory organ that had already a hearing loss for the high frequencies, e.g. as a result of a noise trauma or presbyacusia. Together with the bass deafness, a hearing loss for the high tones may also arise, while the hearing in the middle region is approximately normal or at least better than in the bass region. Figure 1 shows what the audiograms in the various cases approximately look like.

The type I audiogram shows the hearing loss in case of a pure bass deafness. The type II audiogram shows the hearing loss in case of a simultaneous disturbance in the region of the low and the high tones, the middle region being relatively normal. The type III and IV audiograms show a hearing loss in case of a bass deafnass, combined with a presbyacusia and a noise dip respectively.

The terms low frequencies and high frequencies are relative in this connection. The bass deafness may be restricted to the region of the lower tone limit up to and including 250 c.p.s., but may also spread up to and including for instance 2000 c.p.s.

The bass deafness may be unilateral but a bilateral one occurs too.

The perceptive character of it finds expression in the fact that the air conduction curve and the bone conduction curve coincide in the audiogram. When there is a unilateral bass deafness, the bone conduction of the affected ear must in any case be determined while the normal ear is being masked at the same time. It is often recommendable to determine the air conduction too with due observance of this measure.

During Weber's test the tone is lateralized to the nonaffected ear. This is not always the case, for often the patient cannot indicate in which ear the tone is heard best. This finding also tells strongly in favour of the perceptive character of the deafness as the tone, in case of a conduction deafness, is mostly lateralized very easily and quickly to the affected ear.

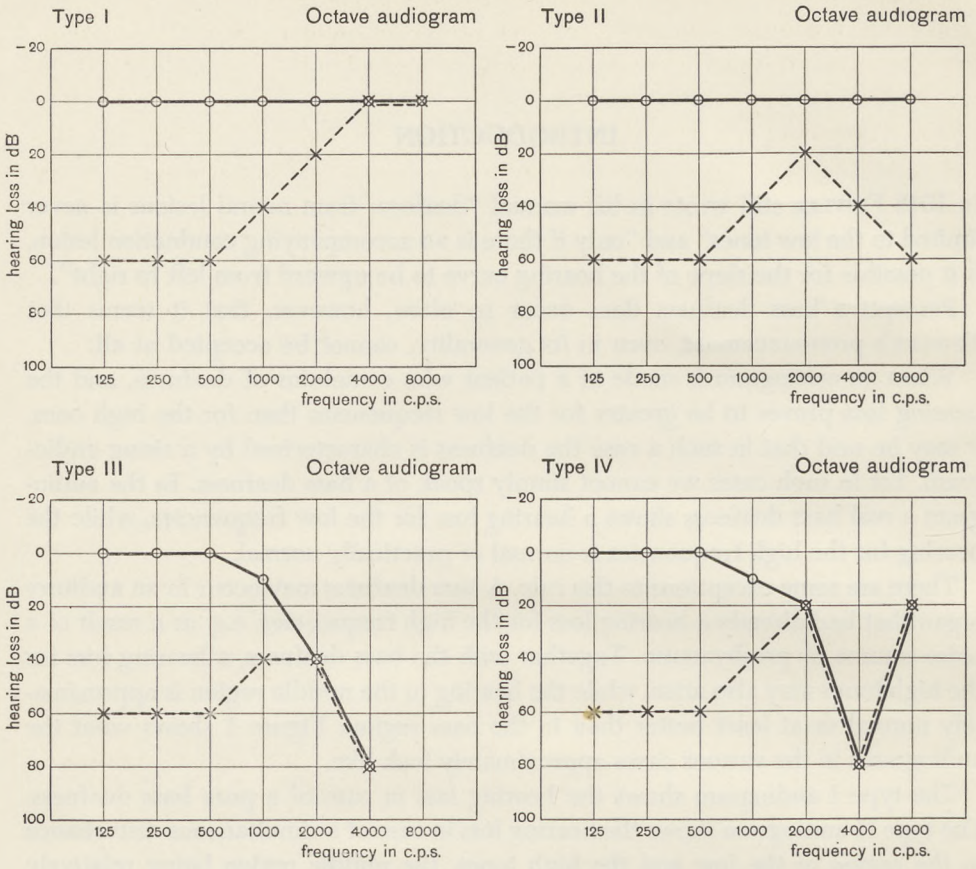


Figure 1

The tests of Schwabach and Rinne should also take place while the nonaffected ear is masked at the same time, as otherwise here is the chance that Schwabach's test proves to be unshortened and Rinne's test negative. If the nonaffected ear is not masked we run the risk that Schwabach's test actually gives us the bone conduction of the nonaffected ear; this conduction then proves to be unshortened. With Rinne's test we compare in that case the bone conduction of the nonaffected ear with the air conduction of the affected ear and Rinne's test seems negative (is then pseudonegative).

What has been described above proves very clearly that in these cases it is essential during the hearing tests to mask the nonaffected ear, as we are otherwise led astray time and again and think we have found a conduction deafness where in fact there is a perceptive deafness. Nilsson pointed out this cause of many errors in 1942. Yet many patients with a perceptive bass deafness are being treated for a tubal catarrh as a result of the fact that the above-mentioned precautionary measure was disregarded while determining the auditory function.

The existence of perceptive bass deafness was pointed out by some investigators

in the course of years. So if there can no longer be any doubt about the occurrence of this form of deafness, there is still no consensus of opinion about the question where the lesion causing this deafness, must be localized in the auditory pathways. Some writers are very positive when they say that the lesion must be found in the cochlea. They point out for instance that bass deafness occurs within the scope of Ménière's disease. Others describe injuries in the central nervous system and they assume a causal connection between the various findings.

We shall endeavour to throw light on this matter in the following chapters.

In *chapter I* we shall give a survey of clinical descriptions that occur in the literature and have a bearing on bass deafness. At the same time we shall pay attention to the explanations given by the various authors of the syndromes described by them.

In *chapter II* we shall discuss experiments made by various investigators for the purpose of causing a bass deafness by means of a cochlear lesion. These experiments were made with animals and a model of a cochlea.

In *chapter III* we shall occupy ourselves with the pathologico-anatomical findings in the mastoids of patients who had suffered from Ménière's disease in their lifetime. The audiograms made before death are also taken into consideration and all this is connected with the fact that it is often assumed that the disturbances in the hearing in cases of Ménière's disease are especially characterized by a bass deafness.

In *chapter IV* experimental and clinical cases of bass deafness are described, based on a lesion of the VIIIth nerve or the more centrally situated auditory pathways. The data obtained from the literature were supplied by patients and experimental animals.

In *chapter V* some instances will be given of the remarkable behaviour of the hearing in the bass region if the VIIIth nerve is operated upon or if the patient is treated with streptomycin.

In *chapter VI* we shall go into the possible mechanism of the origin of bass deafness.

In *chapter VII* the methods of investigation will be described which were applied by us to our own patients.

In *chapter VIII* we shall give the result of our own examination of 45 patients suffering from bass deafness. The case histories of a group of 19 patients will be discussed in detail.

In *chapter IX* the findings in the auditory organ, obtained from our patients, will once more be carefully considered. We shall try to explain the various conflicting results of the examinations. Finally, reverting to chapter VI, we shall try to amplify the hypothesis put forward in that chapter concerning the origin of bass deafness.

Chapter I

CLINICAL OBSERVATIONS ON BASS DEAFNESS

In 1896 Siebenmann described the syndrome, caused by tumours of the corpora quadrigemina region. Besides the numerous neurological symptoms there was impairment of hearing, mostly on both sides and in an early stage limited to a diminished perception of the low tones; later on an equal impairment of hearing occurred for all frequencies.

Krassnig (1924, 1928) made some communications about hearing impairment in affections of the nuclear region of the VIIIth nerve. In 1950 he published a paper on the clinical picture of deafness due to lesions of the brain stem, in which he distinguished between the encephalitic and the traumatic forms. In the first case encephalitic foci in the floor of the IVth ventricle were considered to be the cause of the clinical picture; in the second case a hemorrhage or an embolus were supposed to have occurred. Neurologically bulbar symptoms came to the front. The spoken voice was understood badly, the whispered voice relatively well. The ticking of a watch was heard well. The tuning forks below 2048 c.p.s. were heard badly, those above this frequency were heard well. So here we have a real bass deafness. Krassnig collected about fifteen of these cases. In one case it was possible to make a pathologico-anatomical investigation; this case will be discussed in chapter IV. The author supposed that affections in the floor of the rhomboid fossa will especially damage the pathways of the low and middle frequencies; the pathways for the higher frequencies were thought to have a more superficial course and therefore to be less subject to higher pressure of the tissue. It is a pity that the audiological findings were limited to the results of the tuning fork tests and that no audiograms were made.

Tonndorf (1928) published an observation concerning a 19-year-old girl. The patient complained of deafness in the left ear which had suddenly occurred two years before. In the beginning the deafness was attended with vestibular disturbances, but these had passed off quickly. An otoscopic examination did not reveal any alterations.

The whispered voice was heard ad concham, the spoken voice at 20 cm. The tuning fork tests revealed that 32 c.p.s. was not heard, 64 up to and including 256 c.p.s. only when the fork was struck vigorously, 512 c.p.s. was heard much better; the hearing for 1024 and 2048 c.p.s. was only very little impaired and 4096 c.p.s. was heard normally. The upper tone limit was 17000 c.p.s. The right ear was normal. There was no spontaneous nystagmus and the left labyrinth gave normal responses to calorization. The author based his explanation of the hearing

impairment on the arterial supply of the labyrinth; the apical coils of the cochlea are supplied by the ramus cochlearis propria of the arteria auditiva interna (SIEBENMANN). A hemorrhage or an embolus in this region was considered possible but not likely since the girl was otherwise healthy. A hydrops labyrinthi was thought to be equally improbable in view of the isolated hearing loss for the low frequencies. Finally a vascular spasm was assumed, an "angioneurotic crisis", which, as was generally thought, caused irreversible damage to the labyrinth if this continued long enough.

According to DEDERDING (1932) bass deafness is often met with in cases of brain affection. This, she thought, was caused by an increased intralabyrinthine pressure existing in these cases. The fenestrae thus become fixed and we have in reality an affection of sound conduction and consequently the result is a bass deafness. Pathologico-anatomical investigations have proved that in tumours of the brain the labyrinth very often is markedly choked owing to the intimate connection existing between the vascular and, to a certain extent, also the fluid system of the labyrinth and the cranial cavity. The author derived evidence for her conception from some examples in which hearing kept pace with the symptoms of brain pressure and especially with the development of a choked disk.

MYGIND (1932) shared this way of thinking. In cases of trauma capitis, epilepsia, encephalitis and multiple sclerosis bass deafness may occur as well, according to this author, and it was thought that this was based on a choked labyrinth too.

NYLÉN (1939) stated that there are several reasons for the belief that labyrinthine stasis does not play any important part in cases of tumour of the brain, including histological investigations of human labyrinths, experiments on animals and clinical observations. In his material there were several cases without any signs of stasis in the fundus oculi but with cochlear and vestibular symptoms as pronounced as in the cases with papillary stasis. Furthermore there were several cases with pre-operative papillary stasis, which was relieved by operation, whereas abnormal hearing and abnormal vestibular symptoms remained, and sometimes even increased.

GÜTTICH (1934) proposed to use this conception of the choked labyrinth no more. In cases of an acute rise of the intracranial pressure, passive hyperemia and, because of congestion, hemorrhage and transudation may occur but chronic intracranial hypertension leaves the labyrinth unaltered. The hearing disturbances which occur with tumours of the most different localizations are best explained by pressure on nerve, auditory pathways and centres.

GRAHE (1932) too is of the opinion that in cases of intracranial hypertension there are no characteristic hearing disturbances; the hearing may be impaired in many ways.

This author described the case of a 56-year-old man. Examination revealed an impaired corneal reflex, a hyperesthesia of the Vth nerve, paralysis of the second and third branch of the VIIth nerve, ptosis and enophthalmus, all on the left side; perspiration only on the right side. There was a hearing loss in the left ear for the frequencies 64 up to and including 512 c.p.s.; both ears had a high tone loss.

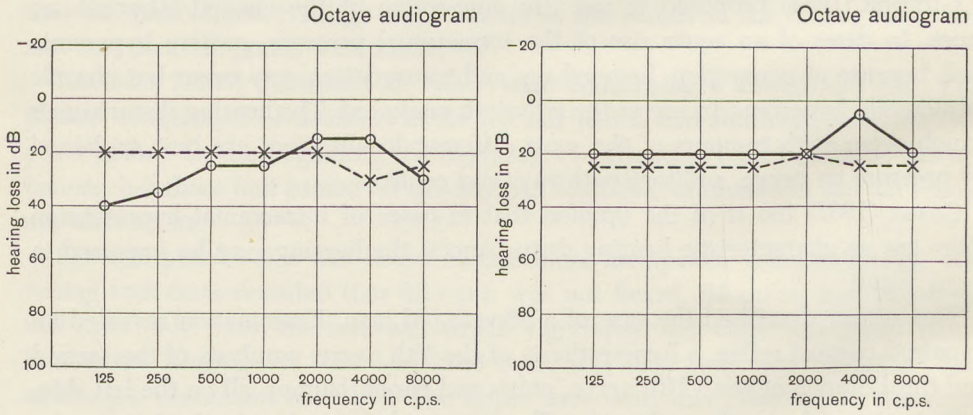
The vestibular tests showed a preponderance of the nystagmus to the right. It was supposed that there was a focus in the medulla oblongata on the left side in the vicinity of the auditory pathways.

In a second patient a choked disk was found. Operatively a tumour was removed from the cerebello-pontine angle on the left side. Before the operation there was a hearing impairment for the high frequencies in the right ear; after the operation the hearing for the high frequencies had improved and a bass deafness had arisen instead.

A third case concerns a 19-year-old patient with the remaining symptoms of an encephalitis that had begun three years before. There were slow pupillary reactions and enophthalmus, an impaired corneal reflex, paresis of the VIIth and XIIth nerves, weakened reflexes of the extremities and the abdomen, adiadochokinesis, hypesthesia of arm and neck, all on the left side. In the right ear there was a hearing loss for the frequencies 64 up to and including 1024 c.p.s. There was a spontaneous nystagmus of the second degree to the left. It was supposed that there was an encephalitic focus in the vicinity of Deiters' nucleus on the left side.

SHAMBAUGH (1935, 1940) described 45 patients with diplacusis. Clinical experience confirmed his theoretical statement that diplacusis may only be caused by a lesion of the organ of Corti. Attacks of vertigo occurred in many of the patients with inner ear deafness and diplacusis so that the cases may be regarded as instances of Ménière's disease. In some cases it was apparently the result of focal infection; allergy was the cause of it in at least some other cases. The loss of hearing for low tones was as great as or greater than that for high tones. The author surmised that the lesion was probably inflammatory in nature and that the vibrating basilar membrane was weighted down by an inflammatory exudate. The following case is representative for his material.

A 57-year-old man complained of dizziness; he was not conscious of tinnitus or deafness but the audiogram showed a bass deafness in the right ear (figure 2).



first examination

3 weeks later

Figure 2

Definite diplacusis was present, the low tones being heard at a higher pitch in the affected ear. A diagnosis of exudative labyrinthitis was made. Careful search for focal infections in the nose and throat gave negative results, but the patient was convalescing from a prostatectomy complicated by cystitis, for which he was

Figure 3

still under treatment. The labyrinthitis was possibly the result of the infection of the bladder. Three weeks later the hearing had improved distinctly. Seven weeks later the hearing was normal and there had been no more dizziness.

WRIGHT (1938) too described some cases of bass deafness. He made a critical analysis of seventy-three cases of aural vertigo. From the evidence he deduced that the labyrinthine disease resulted from a focus of infection, usually situated in nose, throat or mouth. He suggested the name of "focal labyrinthitis" for this disease.

ASHERSON (1940) described an unusual case of polyneuritis, which he termed a "polyneuritis of the porus acusticus". The facial, the vestibular and the cochlear nerve were affected. The paralysis was transient and complete recovery followed. The paralytic wave swept across both the VIIth and the VIIIth cranial nerves in the following chronological order: vestibular nerve – facial nerve – high tone branch of the cochlear nerve – low tone branch of the cochlear nerve. By the time the last branches were involved, resolution was already apparent in the parts first affected. Recovery was in a reverse order; the entire process lasted one month (figure 3).

WALKER (1942) and SALTZMAN (1952) studied the hearing loss in patients who underwent stereoencephalotomy. Some of their cases will be described in chapter IV.

LINDSAY (1944, 1946) described three cases of Ménière's disease, two of them having a distinct hearing loss in the bass region. His material will be discussed in chapter III.

SCHUKNECHT (1950) published the case of a 25-year-old student with a trauma capitis. The patient was unconscious for some hours; there was no bleeding from the ears. On the radiograph a fracture of the skull was seen, but this did not pass through the temporal bone. Some days later a leftsided deafness was noted. There were also complaints of a slight dizziness, of the type frequently seen after a commotio cerebri. The ear drums showed no abnormalities. At calorization the left labyrinth was less excitable than the right. In the Weber's test the tones were

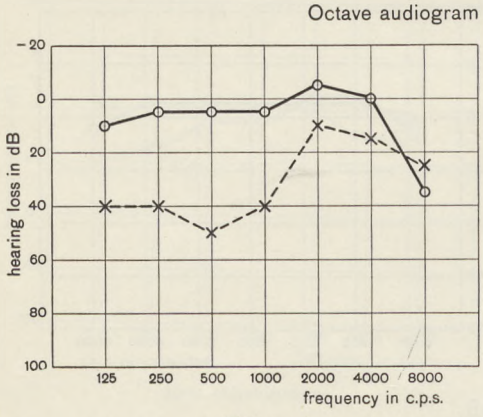


Figure 4

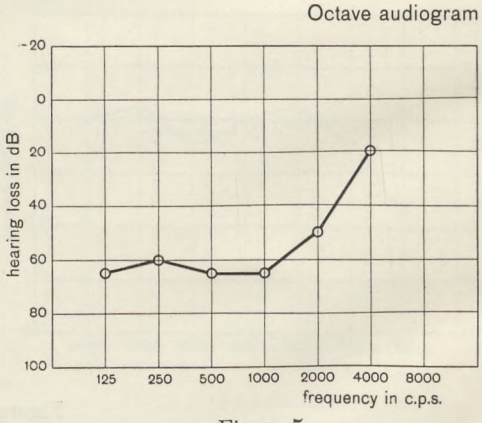


Figure 5

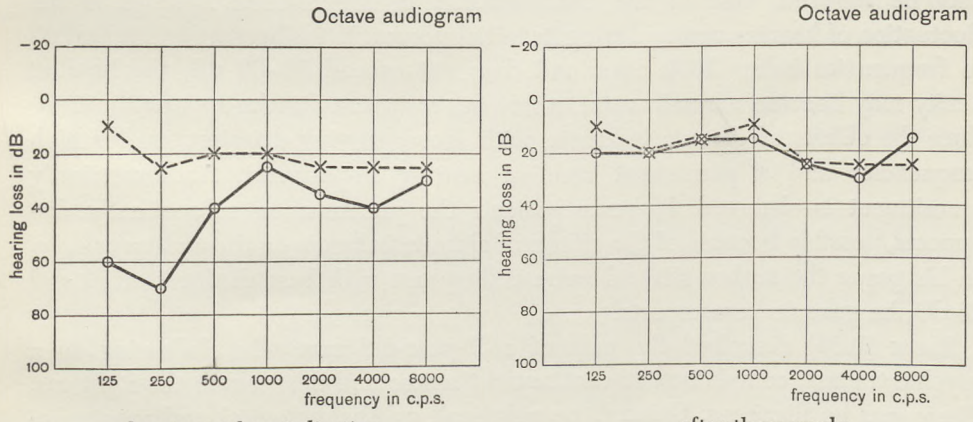
lateralized to the right. The audiogram showed a bass deafness in the left ear (figure 4), characterized by the presence of the recruitment phenomenon. The possibility of a fresh case of Ménière's disease was considered; the patient did not complain of tinnitus.

WILLIAMS (1952) considered the physiological basis of allergy of the inner ear, usually termed Ménière's disease, in its several variations. There are autonomic dysfunctions with their stereotyped or invariable reactions on the peripheral vascular bed, resulting in transudation of an excess fluid high in protein content and consequent elevated osmotic pressure. The vestibular symptoms seem to be mainly due to vasospasms, while the cochlear symptoms appear to depend mainly on an increase in endolymph of high osmotic pressure, according to this author. One of his cases will be described here.

A 38-year-old man complained of attacks of vertigo. There was an increasing deafness. The threshold audiogram for pure tones showed normal findings on the left. On the right there was a distinct bass deafness (figure 5). Loudness balance tests showed an incomplete recruitment of loudness in the right ear over that of the left at frequencies of 500, 1000 and 2000 c.p.s. The equilibrial function of the right labyrinth was relatively decreased. A roentgenogram of the skull showed evidence of Paget's disease.

GREINER and his collaborators (1953, 1954, 1956) described the occurrence of bass deafness in the course of affections of the central nervous system. Two of their cases will be discussed here, two other cases are to be described in chapter IV.

A 45-year-old man was hospitalized with the following symptoms. There was a third degree rotatory nystagmus to the left. Conjugate deviation of head and eyes to the right. Rightsided facial paresis. Rightsided facial hypesthesia with weakened corneal reflex of the same side. Hiccough. Impossibility of swallowing. The uvula was distorted to the right. Stagnation of saliva in the valleculae. Absence of contractions of the pharynx muscles. Pyramidal disorders with a Babinski on



hearing at hospitalization — after three weeks

Figure 6

the left; hypesthesia of the same side of the body. Disturbances in coordination. So we are dealing with an alternating syndrome due to an affection of the brain stem wich can be localized very well. In addition there was a hearing loss for the low frequencies on the right side (figure 6), attended with recruitment. In three weeks there was a general amelioration; the hearing had improved remarkably.

A 54-year-old man was suffering from attacks of vertigo. There was tinnitus in the right ear and the hearing in this ear was reduced (figure 7). Examination revealed important trophic alterations of the hands. All the signs of a typical syringomyelia were found, localized in the upper extremities, and most marked on the right. On the right side there was a Horner's syndrome and a slight hypesthesia of the right side of the face was found.

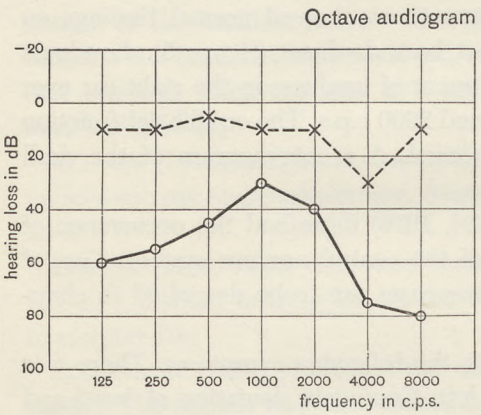


Figure 7

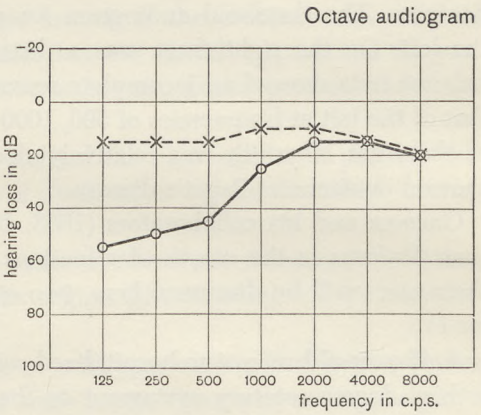


Figure 8

According to Kos (1955) the hearing loss in cases of labyrinthine hydrops is of the inner ear type and usually unilateral, and in the early stage of the disease mainly the low tones are affected. Eventually the hearing for all frequencies may be reduced. One of the characteristics of labyrinthine hydrops is the fluctuation of hearing acuity during its initial phases. If the hearing loss is limited to frequencies below 1000 c.p.s. and does not exceed 20–30 dB, the hearing acuity may become normal again; otherwise, recurrent attacks apparently introduce the effects of chronic ischemia, with accompanying deterioration for high frequencies and a permanent residual hearing impairment. Audiometrically recruitment is observed. In some patients the phenomenon subsequently disappears, possibly because of the effects of chronic ischemia on the auditory nerve. In his paper the author showed some audiograms with bass deafness of 50 and 60 dB; he gave no case reports.

Baum (1955) described the case of a 39-year-old man who, following on a psychic trauma, complained of continuous tinnitus and deafness in the right ear. There was no dizziness. General, neurological, ophthalmological and roentgenological examinations revealed no abnormalities, except a distinct neurovegetative

dystonia with dermographism, tremor of the hands, etc. Vestibular tests revealed normal responses of both labyrinths. The audiogram showed normal hearing for the left ear; the right ear suffered a hearing loss for the low frequencies (figure 8) with complete recruitment (Fowler's test) and lowered differential threshold (Lüscher's test). The noise-audiograms (Langenbeck) were of the inner ear or hair cell type. Based on these findings the author assumed an inner ear hair cell lesion in the apex of the cochlea near the helicotrema. A definite diagnosis could not be established.

Lundborg (1955) published eight out of about thirty cases of bass deafness in which a thorough, topographical diagnosis was made and in which investigation showed various types of lesions in the auditory pathways. One of his cases will be discussed in chapter IV. Some other cases will now be briefly revieved. Since the author's audiograms were taken with a Békésy audiometer, they will be translated into octave audiograms.

A 33-year-old woman showed acute symptoms of partial facial nerve paresis and neuralgic pain in the trigeminal region on the right side, and subsequent left-sided trigeminal neuralgia, loss of hearing and dizziness. Objective examination revealed a left-sided nerve-type hearing loss for low tones (figure 9). Further clinical and neurological investigations, including encephalography, gave negative results. These symptoms of both the facial and trigeminal nerves of the right side and the trigeminal, cochlear and vestibular nerves of the left side were interpreted as a kind of encephalitis.

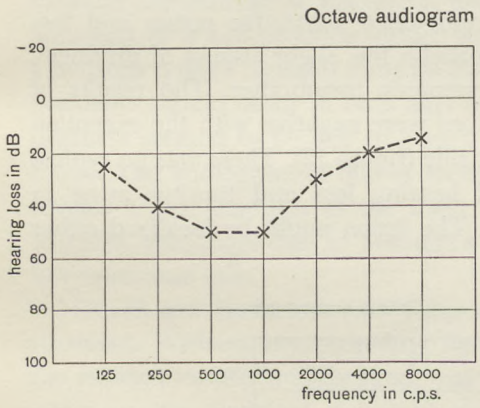


Figure 9

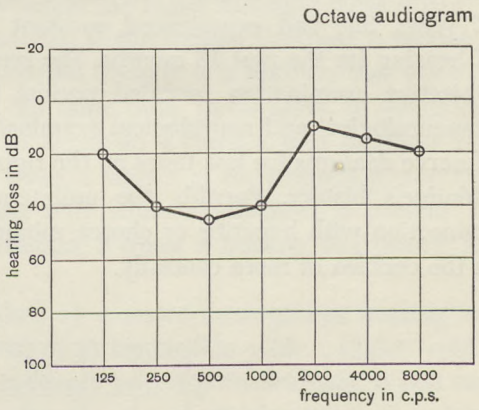
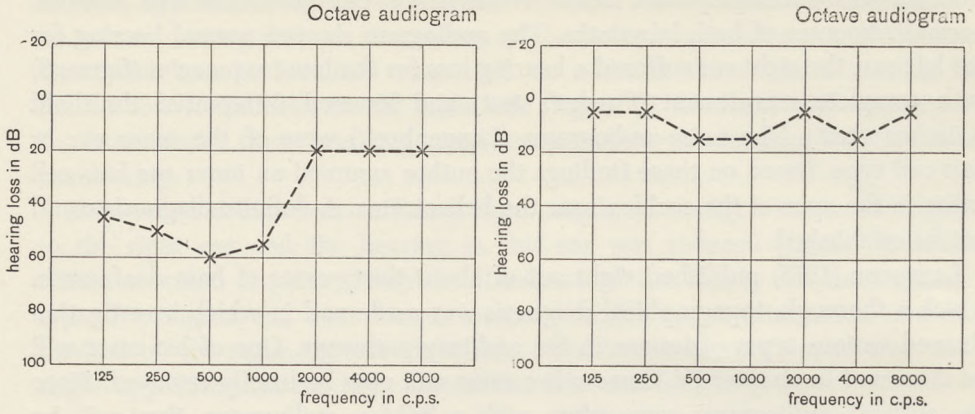


Figure 10

A 35-year-old woman with an earlier history of polyarthritis and hepatitis, and subsequent persistent periodic headaches in the frontal region, reported left-sided loss of hearing of several years' duration. A thorough, clinical investigation including a neurological examination and porus roentgenograms revealed no pathological condition other than left-sided bass deafness. No diagnosis was established; possibly hepatitis may have been the cause. A follow-up examination six months later showed normal hearing (figure 11). The diagnosis was cephalalgia (encephalopathia infectiosa?).



first examination 6 months later

Figure 11

A 47-year-old woman had a history of Ménière's disease of 15 years' duration, first on the left side. During the last year she had suffered from loss of hearing and tinnitus on the right side as well (figure 10). The patient underwent a thorough neurological examination. She was operated on (division of the vestibular nerve), on the left side one year after the appearance of symptoms. Her dizziness disappeared after that. The patient's hearing and tinnitus remained unchanged.

A 37-year-old woman had a history of hepatitis and chorea minor in childhood. She was otherwise healthy and there had been no ear symptoms for the past 10 years. She had experienced constant right-sided subjective noises and loss of hearing for the past 18 months. She reported a few slight attacks of dizziness. Objective examination revealed normal tympanic membranes. The results of otoneurological and neurological examinations were negative with the exception of nerve deafness for low tones on the right side (figure 12). There was no typical Ménière's history. Possibly the unilateral hearing loss and tinnitus arose in connection with hepatitis or chorea minor. The lesion might be localized either in the cochlea or more centrally.

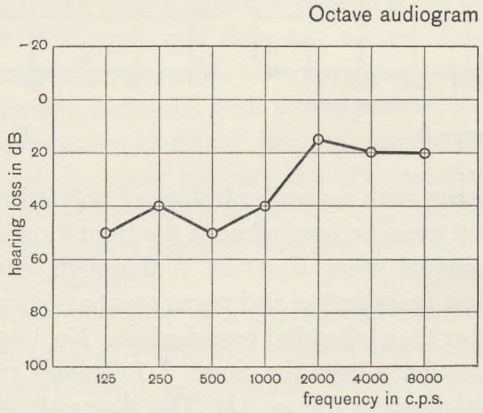


Figure 12

According to Lundborg the cause in some cases of bass deafness is unknown after investigation; in others lesions of the central auditory pathways (encephalitis, acoustic tumours) as well as labyrinthine lesions (Ménière's disease etc.) have been found. Békésy audiograms without signs indicating cochlear damage (recruitment) and a negative nerve status both indicate localization in the auditory nerve itself (acoustic neuritis). Nerve deafness for low tones does not indicate any special lesion from the point of view of topographical diagnosis.

Walsh (1956) pointed out that, although the patient complains of vertigo in cases of Ménière's disease, he gives the information that he has difficulty in hearing. The usual complaint is one of fullness in the affected ear, which usually becomes worse just before an attack of vertigo. The patient notices a great fluctuation in his ability to hear and says that sometimes he can "hear perfectly" and at another time is "quite deaf". In the early stages of the disease the pure tone audiogram shows a greater loss in the low than in the high tones. As the disease progresses the audiometric curve tends to flatten and the loss may be equal throughout the scale. Later in the disease there is usually an increasing loss in the high tones. Characteristically there is a much greater discrimination loss for speech than would be suspected from the pure tone audiogram. While the threshold loss for speech may be no greater than 40 dB, the discrimination score is usually less than 50 % and is quite often below 30 %.

Hallberg (1956) wrote a paper on sudden deafness of obscure origin. According to this author the vascular supply of the labyrinth is such that a thrombus, vascular hemorrhage or vasospasm may affect the entire auditory sense organ or only part of it and produce sudden deafness. If the sudden deafness is bilateral, a hydrops type of disorder must be considered because it is unlikely that vascular accidents should occur in both ears at the same time. In 89 cases, in which the deafness seemed to be due to a vascular accident, 4 times a bass deafness was found. The audiogram of one of these cases is shown in the original paper; this reveals a bass deafness of the inner ear type while recruitment is present. In 56 cases, seemingly due to Ménière's disease, 5 times a hearing loss mainly for low tones was seen.

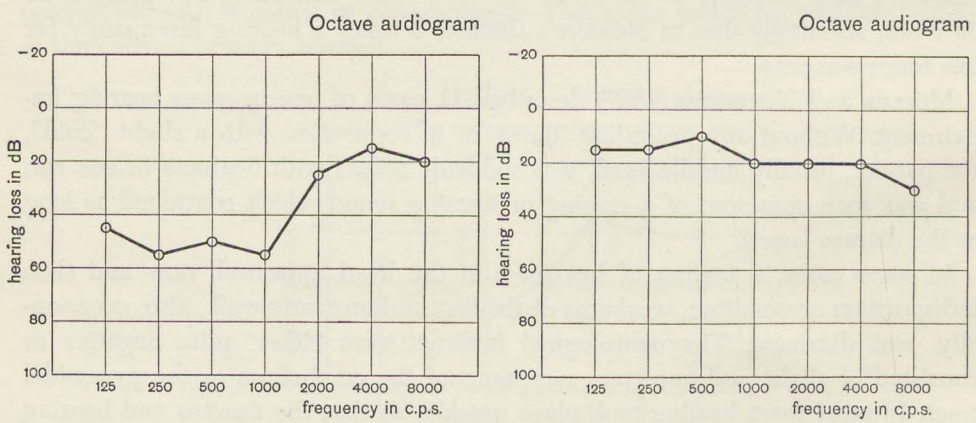
Möller and Nenzelius (1957) described 11 cases of neurogenous hearing impairment. Without any preceding illness, or in connection with a slight "cold", the patient, usually middle-aged, was suddenly seized with deafness in one ear, and was soon conscious of a ringing or buzzing sound which continued as long as the disease lasted.

In some cases, a feeling of heaviness in the head appeared, now and then indisposition or vomiting, weakness of the legs and unsteady walk, also, occasionally, real dizziness. The neurological findings were either quite negative or consisted of slight and uncertain symptoms of the cerebellum or the pyramidal tract. In most cases healing took place quickly, so that the tinnitus and hearing impairment had entirely or practically disappeared after three weeks. The authors presumed that this cochlear paresis was a form of brain stem encephalitis, referring either to an inflammatory or possibly to an allergic process. One of their cases

showed a real bass deafness of 35 dB without recruitment (Békésy method).

According to Opheim and Flottorp (1957) reduced hearing and tinnitus are regular symptoms in Ménière's disease, and may be very marked, partly dominating the picture and so obscuring the vertigo. For long periods they may even constitute the only clinical manifestations of the disease. The hearing loss frequently manifests itself predominantly within the low frequency range of the auditory field below 2000 and 3000 c.p.s. The hearing loss frequently presents considerable fluctuations, generally also in the low frequency range below 2000–3000 c.p.s. These fluctuations even occur after a duration of the disease of several years, and necessitate repeated examinations as they may otherwise be overlooked. As long as the hearing loss has not reached such a degree as to approach total deafness, it is consistently characterized by the presence of the recruitment phenomenon, demonstrated by a positive Fowler's test and/or low aural harmonic thresholds. Intolerance and disturbed pitch perception also occur very frequently. These distinctive features confirm that the site of the disease is peripherally in the labyrinth. The fluctuating low frequency hearing loss together with the recruitment phenomenon, intolerance and disturbed pitch perception, constitute a symptom complex of a high diagnostic value.

One of the case reports of these authors will be reproduced here. It concerns a 45-year-old man whose hearing of the right ear had not been quite so good for the past few years. He had suffered from several attacks of gyratory vertigo. Tinnitus in the right ear was persistenly present. The audiogram showed a bass deafness of 50 dB in the right ear. The hearing of the left ear was normal (figure 13). In the right ear the hearing loss was combined with abnormally low aural harmonic thresholds, reduced tolerance, binaural diplacusis and a positive Fowler's test. Four months later approximately normal hearing was found in the right ear for the low frequency range, a normal harmonic threshold, no intolerance and no binaural diplacusis.



first examination

4 months later

Figure 13

At present it is no longer possible to judge the results of tuning fork tests as published by earlier investigators according to their real merits.

The publications by Siebenmann, Krassnig, Tonndorf, Dederding, Mygind, Nylén, Güttich, Grahe and Wright were discussed for the sake of completeness, as they all found a more or less pronounced form of bass deafness in their patients by means of the tuning fork test. It is by no means certain that an audiometrical examination of their cases would have led to the same conclusions. Nevertheless it is interesting to observe that the most different kinds of affections were already held responsible for the origin of bass deafness. We may mention here tumours of the corpora quadrigemina region (Siebenmann); encephalitis, hemorrhage or tumour in the brain stem (Krassnig, Grahe); tumour of the brain in general (Dederding, Mygind, Nylén, Güttich), which presented a point at issue whether hearing impairment was caused by a choked labyrinth or by pressure of the tumour on auditory pathways or centres. We may add to this hemorrhage, embolus or vascular spasm in the apical coil of the cochlea which is supplied by a terminal artery, the ramus cochlearis propria of the arteria auditiva interna (Tonndorf) and finally foci of infection in nose, throat and mouth, which lead to a "focal labyrinthitis" (Wright).

Owing to the fact that the audiometer, a device to record any loss of hearing for every frequency in exact units and numbers, came into general use somewhere about 1940, the publications that have appeared since that time can be more easily discussed.

Here too authors mention various affections which might cause bass deafness: labyrinthitis (Shambaugh); neuritis of the VIIIth nerve (Asherson); trauma capitis (Schuknecht); allergy of the inner ear (Williams); affections of the brain stem (Greiner, Lundborg, Möller and Nenzelius); labyrinthine hydrops (Kos); inner ear hair cell lesion in the apical coil of the cochlea (Baum), Ménière's disease (Walsh, Hallberg, Opheim and Flottorp); disturbances in the vascular supply of the labyrinth (Hallberg).

Lundborg's pronouncement, that bass deafness is not typical for a certain affection from the point of view of topographical diagnosis, is obvious in every respect. On the other hand we must admit that quite often a diagnosis was established on little more than hypothetical grounds.

So there is some sense in studying closely those cases of bass deafness, in which a lesion in cochlea or auditory pathways could incontestably be proved. Suitable material consists of patients who showed a disturbance in the normal function of the auditory organ owing to a pathological process or to an operation, and in whom place and nature of the lesion could be determined, either by a microscopical examination after death or during the operation. At the same time we can avail ourselves of experimental animals by deliberately causing damage to cochlea or auditory pathways while determining the acuity of hearing before and after the interference. A microscopical examination after death may complete our data in these cases.

Chapter II

BASS DEAFNESS DUE TO COCHLEAR LESIONS

In 1920 Matsui experimentally tried to cause hemorrhages in the labyrinths of animals, by means of congestion of the head. It is true that in these experiments with guinea pigs no attention was paid to the effects on the hearing, but they are worth mentioning in this connection. Congestion of the head was achieved in several ways. In some animals the cervical sympathetic trunk on both sides was cut. The labyrinths of these animals showed dilated vessels, a slight bleeding in the apical coil of the scala tympani and only insignificant changes in the hair cells of the organ of Corti and in the ganglion cells. In other animals congestion of the head was achieved by a cuff around the neck; this method caused significant changes. The labyrinths showed dilatation of the vessels in the spiral ligament. Hemorrhage in the cochlea, especially in the apical coils, was found. The hair cells of the organ of Corti showed obvious pathological changes. The ganglion cells and the nerve fibres were degenerated too. Matsui stressed the fact that in these circulatory disturbances essentially the apical coils of the cochlea were affected.

Davis, Lurie and Stevens (1935) studied the electrical potentials generated in the cochleas af guinea pigs; these potentials were picked up by electrodes placed on the round window and the neck. Electrical audiograms were made before and after drilling through the cochlear wall and damaging the organ of Corti at various levels. The exact location and extent of each lesion was determined by subsequent microscopic study of serial sections of the cochleas and these were correlated with the losses in hearing revealed by the audiograms. The losses in most cases were not greater than 20 to 30 dB. They found that high tones are localized near the basal end of the cochlea; 2000 c.p.s. is in the middle and the low tones are gathered in a bunch quite close to the helicotrema. A tone of 20 or 30 dB above the threshold apparently activates a considerable zone of the organ of Corti, but this fact does not in any way contradict the fundamental localization described above.

Hughson, Thompson and Witting (1935) used cats for their experiments. The electrical audiograms were obtained with the Wever and Bray technique – i.e. by using a nerve electrode. Small holes were drilled in the wall of the cochlea at selected levels from the base to the apex. The base of the drill hole was then scorched with a high frequency cautery. This experimental damage to the organ of Corti caused a loss of transmission for the higher frequencies. Dependent on the extent of the lesion, no matter how this was caused, the higher frequencies

were first lost. This fact held true regardless of the site of the lesion. Extensive lesions could be caused at the apex without losing any frequency from 64 tot 4096 c.p.s. The invariable loss of the higher frequencies in the presence of greater damage to the organ of Corti at any point, suggests a quantitative relationship between the amount of organ of Corti present and the perceptible frequency range.

Bast and Eyster (1935) measured the electrical potentials generated in the cochleas of guinea pigs under various conditions. They found that low frequencies can be picked up more readily from the apex than from the base, while high frequencies are picked up more readily from the base than from the apex. Removal of the apical turn and then removal of the second and third apical turns produced a marked reduction of response to the low frequencies, while the higher frequencies were only slightly reduced. But their study also showed that normal transmission curves can be obtained in the guinea pig when there is complete atrophy of the organ of Corti in all turns. It is apparent then, according to these authors, that the integrity of the organ of Corti is not essential to the production of cochlear potentials and that therefore these potentials are probably not an index of the actual hearing of the animal. The cochlear potentials, however, while not dependent on the integrity of the organ of Corti, may very well depend on the integrity of other structures that are intimately related and necessary to hearing. By puncturing the cochlear canals, they showed that considerable differences in relative pressure between endolymph and perilymph greatly reduced or nullified the response.

Culler (1935) confirmed the findings of Bast and Eyster. He found that each frequency within the audible range has its own focus of response within the cochlea. This focus is revealed by the electric potentials, which can be shown to be maximal for a single frequency at a given site.

Walzl (1939) studied the effect of local application of salts to the cochlea on cochlear potentials. It was shown that when crystals of NaCl were put in the scala vestibuli of the apex, the first effect was a rapid impairment for the low tones. This impairment did not exceed 25 dB for 256 c.p.s. Later on there was impairment for all tones.

Walzl and Bordley (1942) studied again the effects of localized lesions of the cat's organ of Corti on the thresholds of cochlear potentials. Histological examination showed that the lesions were restricted to the organ of Corti, and that both Reissner's and basilar membranes had remained intact. Reconstructions were made to determine the exact location of each lesion. Single small lesions resulted in a moderate impairment of response (about 20 dB), which was limited to a small part of the frequency range. Larger lesions caused a greater degree of impairment and affected a wider part of the tonal range than the small ones. The authors conclude that near the threshold the response to any frequency is localized to a small part of the organ of Corti (high tones being towards the base), but that with the increase in intensity there is a spread of the response to adjacent areas, the spread being greater for low than for high tones.

Tasaki and Fernández (1952) developed techniques to record cochlear responses from different turns of the guinea pig cochlea simultaneously. They perfused the cochlea with chemicals and polarized the sensory endings in the cochlea by means of direct currents. Complete replacement of the perilymph by normal mammalian Ringer's solution does not cause any change in the electrical responses of the cochlea to sound stimuli. An increase in the KCl content of the perfusing fluid reduces the cochlear microphonics reversibly. Elimination of the responses of the apical turn by an isotonic KCl solution does not affect the responses of the basal turn. Complete suppression of the responses of the basal turn by KCl or a direct current does not influence the cochlear microphonics of the third turn. The basal turn of the cochlea responds to high, to middle and to low-frequency sounds. The third and apical turns respond only to low-frequency sounds. A direct current traversing the cochlear partition from the scala vestibuli (source) to the scala tympani (sink) enhances the cochlear responses. A current flowing in the opposite direction brings about depression of the responses.

It is true that the measurements of the cochlear potentials have furnished many valuable data relative to tone localization in the cochlea, but they can by no means give reliable information about the actual hearing of the test animal. For this purpose the conditioned response method is pre-eminently suitable.

Gross (1952) obtained audiograms by means of the conditioned response method from guinea pigs before and after injury to one cochlea and destruction of the other, in order to study the effects of such lesions on the auditory acuity of these animals to low tones. Injury to the organ of Corti was accomplished either by piercing the cochlear wall with a drill or by application of diathermy to a localized point in the cochlear wall. The postoperative thresholds of three animals were unstable, showing impairment which progressed with time. In seven animals the postoperative thresholds remained stable over a long period of time. The first postoperative tests disclosed that the greatest loss was always obtained for the lowest test frequency and least impairment was always obtained for the highest tone. In figure 14 the postoperative audiograms of nine animals are given; in each threshold curve the extent of the destruction of the organ of

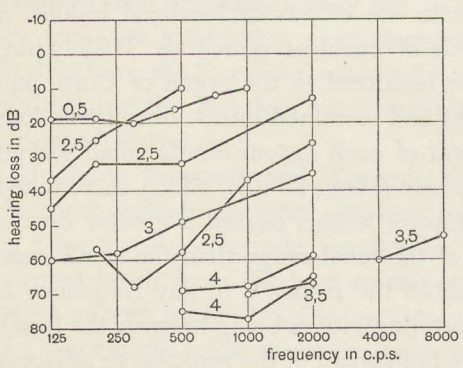


Figure 14

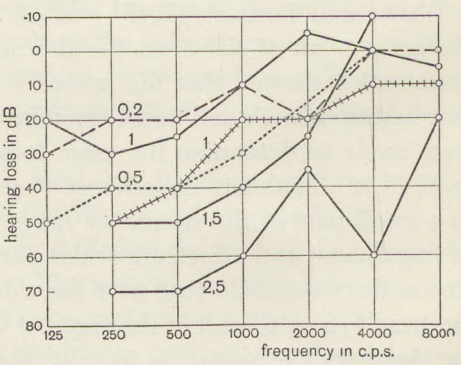


Figure 15

Corti measured from the apex of the cochlea and expressed in the number of turns is recorded.

It is clear that with the destruction of an increasingly larger area of the organ of Corti, as measured from the apex of the cochlea, there is a continued contraction of the range of frequencies to which the animals will respond. This involves the elimination of response to more and more of the lower test frequencies, increasingly greater impairment for the low tones to which the animal will respond and increasingly more impairment for the higher test tones.

The data obtained from this experiment also show that at high intensities, frequencies are able to activate rather widespread areas of the organ of Corti. Animals were able to respond to tones in spite of the fact that the area of the normal organ of Corti lay more than 2½ turns away from the areas of maximal sensitivity for these tones, as disclosed by the electrical recording technique.

SCHUKNECHT and NEFF (1952) developed a surgical procedure by which restricted damage could be done to structures within the apical turn of a cat's cochlea. Audiograms were determined by means of the conditioned response method before and after the production of the intracochlear lesions. After the postoperative tests were completed, the animals were perfused and the experimental ears removed and prepared for histological examination. The cochleas were sectioned serially and graphic recontructions were made. The results indicate that, at near threshold intensities, frequencies below 500 c.p.s. excite receptors only in the apical turn of the cochlea. As the intensity is increased, the low frequencies appear to arouse activity over a wide area of the cochlea so that, at the intensities 20 to 30 dB above the threshold, they are producing excitation at least as far towards the base as the lower middle turn of the cochlea. Because of the difficulty of defining precisely functional and non-functional receptor cells on the basis of histological examination, the results must be interpreted with some caution; nevertheless the following two observations are of very great interest.

In one cat the cochlear injury resulted in a loss of outer hair cells throughout the upper 12 mm. of the cochlea with partial loss of nerve fibres and ganglion cells in the same region. The postoperative threshold curve showed a hearing loss of approximately 50 dB for all frequencies below 2000 c.p.s.; at 4000 c.p.s. and above the deficit was much less. The hearing for low tones can in this case be attributed either to activation of the inner hair cells in the upper turns or to a spread of stimulation so as to affect the outer hair cells in the basal turn.

In a second cat histological examination showed a loss of outer hair cells throughout the cochlea with a spotty loss of inner hair cells in the apical turn and in the lower part of the basal turn.

Nerve fibres and ganglion cells were severely injured in the apical region but were essentially normal in the remainder of the cochlea. A pronounced loss of hearing occurred throughout the frequency range tested: a 55 dB loss for frequencies up to 2000 c.p.s., a 78 dB loss at 4000 c.p.s. and a complete loss for the maximal intensities which could be produced at 8000 and 16.000 c.p.s.

The findings in these two cases (figure 16) might be interpreted thus, that for frequencies in the range below 2000 c.p.s. intensity must be raised 50 to 60 dB above the normal threshold level, before the inner hair cells are stimulated sufficiently to produce neural excitation. Perhaps at intensities above 50 dB the lower frequencies are exciting outer hair cells in the basal turn as well as the inner hair cells in the middle and apical turns.

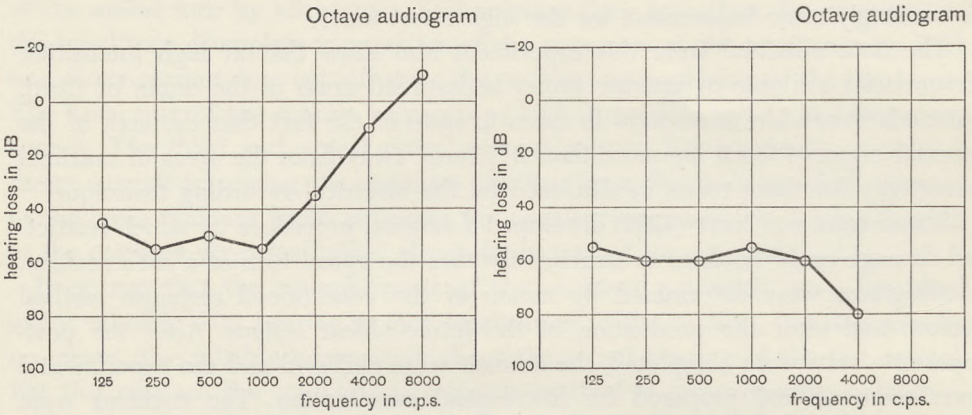


Figure 16

Sutton and Schuknecht (1954) created small cochlear lesions by mechanical injury with fine needles or with controlled electrical currents in the apical turn of the cat's cochlea. The cortical technique was used as a method of evaluating the function of the injured cochlea. For that purpose the auditory cortex was covered with small patches of filter paper soaked in a saturated solution of strychnine sulfate. Monopolar recording took place at each patch in turn. The criterion of cortical response to sound stimuli introduced into the experimental ear was any constant deflection on the oscilloscope, which occurred at a constant latency following the automatically keyed stimulus. The outcomings of this method are in perfect agreement with the results obtained by means of the conditioned response method. In figure 15 the audiograms of 6 cats are given; in each threshold curve the extent of the destruction of the organ of Corti measured from the apex of the cochlea and expressed in the number of turns is recorded.

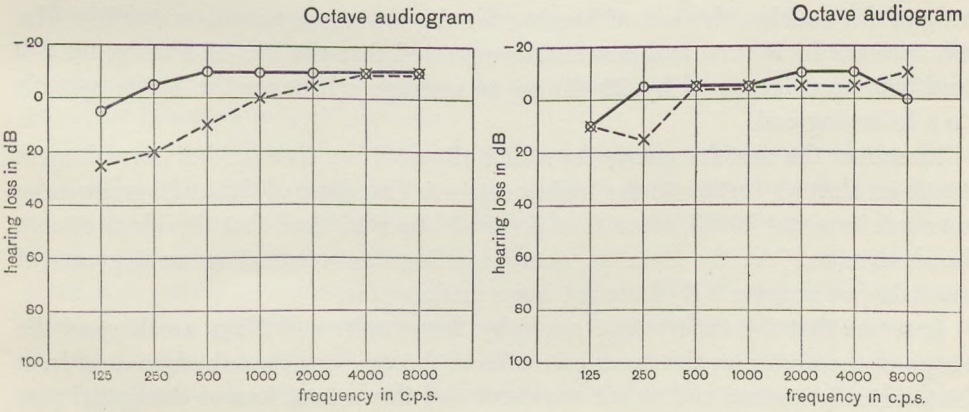
Kimura, Schuknecht and Sutton (1956) confirmed the results previously obtained by Sutton and Schuknecht.

In connection with this chapter the work of Tonndorf (1957) is also of importance. During the years following the first report of Hallpike and Cairns on the histological findings in two cases of Ménière's disease, attempts have been made to explain the hearing loss in this disease on the basis of the hydrops phenomenon. Speculation was focused upon the possibility of a mechanical impairment of sound transmission induced by the hydrops. The rationale for the above may be supported as follows. A distension of the endolymphatic duct stretches its

membranous boundaries. Reissner's membrane, because of its lesser stiffness, bulges considerably, whereas the displacement of the basilar membrane, because of its greater stiffness, is probably so small that it escapes detection during histological examination. In its normal state, the basilar membrane does not possess any tension. Its stiffness is not uniform but varies over its length exponentially; the value at the basilar end being about a hundred times greater than at the apical end. These elastic properties of the basilar membrane favour the formation of travelling waves along this membrane. It is recognized that the travelling waves form the essential link in the transmission of vibratory energy from the stapes footplate to the basilar membrane on its way to the organ of Corti. It was proposed that the alteration of the elastic properties of the cochlear partition, due to distension of the membranes, might affect the pattern of travelling waves within the cochlea. To study this problem Tonndorf used a cochlear model in which he could alter the endolymphatic pressure. It turned out that upon increasing the endolymphatic pressure, the loss per frequency is proportional to the distension of that section of the basilar membrane in which maximal amplitude of displacement is formed for that particular frequency.

The loss is greatest for the lowest tones, the basilar membrane bulging more in the apical than in the basal region. The "hearing loss" curve has a slope of about 1,5 dB per octave in the model case. There is also a shift of the location of maximal displacement of amplitude of the basilar membrane to the higher frequencies upon increase of the endolymphatic fluid pressure (diplacusis).

In his paper the author gives a description of a representative clinical case. It concerns a 32-year-old physician who fell ill with a sudden attack of moderate vertigo and a roaring-type of tinnitus in his left ear. He also noted some impairment of hearing acuity in the same ear. One day after the onset of the symptoms an audiogram was taken (figure 17). Recruitment was not present. Slight diplacusis was noted for some of the lower frequencies. Two days later the hearing had returned to approximately where it had been before the onset of the illness. This attack was the first the patient had ever experienced.



first examination Figure 17 2 days later

Apparenly it is possible to reduce the hearing acuity for low tones by means of mechanical injury of the apical coils of the cochlea (GROSS, SCHUKNECHT, NEFF, SUTTON, ALEXANDER and his associates, KIMURA).

Congestion of the head may lead, at least under experimental circumstances, to hemorrhages in the apical coils of the cochlea, with attendant degeneration of the organ of Corti and the nerve fibres there (MATSUI). The supposition that a bass deafness might result from this, is quite obvious.

An increase of the pressure in the endolymphatic system leads to a reduced elasticity and consequently to a lower amplitude of the basilar membrane, which is most pronounced in the apical coils of the cochlea. This was at least found in a cochlear model (TONNDORF).

It may be expected that for the hearing the consequences of an impairment of the apical coils of the cochlea with degeneration of the organ of Corti and also the nerve fibres there, will be quite different from those of an increase in endolymphatic pressure in which the organ of Corti and the nerve fibres remain quite unimpaired and only the possibilities of vibration of the basilar membrane are limited.

In the first case part of the organ of perception is irreparably lost and the loss of hearing is not reversible. The low tones which used to be perceived by this part are only heard if they are of a rather high intensity. This might be explained from the fact that low frequencies only cause a localized vibration of the basilar membrane in case of very low intensities; in case of higher intensities the whole membrane is caused to vibrate, so that also the hair cells may be excited which lie in the healthy zone of the cochlea.

Another possibility is that actually the low tones themselves are not heard but their harmonics which may arise in the audiometer apparatus or in the auditory organ owing to distortion caused by alinearity. We shall return to this in chapter IX.

In general we may say that when the apical lesion spreads towards the basal side of the cochlea, the loss of hearing for the low tones increases steadily. The experiments by SUTTON and SCHUKNECHT revealed that the loss of hearing for 250 and 500 c.p.s. increased by 20 dB on an average whenever the lesion spreads to a following coil.

Moreover the next frequency for which the auditory threshold is normal again keeps on shifting further to the higher octaves. The slope of the audiogram curve amounts to about 10 dB per octave. It should be remarked that this slope cannot be determined exactly from an octave audiogram; continuous audiograms of animals (see chapter VII) have not been made so far.

It seems that the experiments made by SCHUKNECHT and NEFF are beyond the scope of the above, as the inner hair cells of the two cats described were at least histologically normal and so we could not call this a total loss of the apical part of the organ of perception. The audiogram of the first cat, however, does not

show any essential difference with the audiograms of the experimental animals of SUTTON and SCHUKNECHT, in which, besides the outer hair cells, also the inner hair cells in the apical coils had been destroyed.

The damage caused in the experiments described, were of course not restricted to the organ of Corti and the nerve fibres; also other parts of the membranous cochlea were sometimes damaged. So it is practically impossible to establish to what extent the damage to the various structures in the cochlea has contributed to the genesis of the loss of hearing that was ultimately found. Such an extensive mechanical damage will only very seldom occur in man.

Many investigators have brought to the fore that a disturbance in the blood supply of the apical coil of the cochlea may cause a bass deafness. The organ of Corti, however, has no blood vessels of its own, the hair cells being supplied from the labyrinthine fluid surrounding them. This fluid circulates and there is diffusion of nutritious substances. Therefore it is hardly conceivable that a disturbance in the blood circulation would lead to a total loss of the apical part of the organ of perception while the adjacent part would keep on functioning in a perfectly normal way. We may rather expect a very gradual transition from the area in which all hair cells have been destroyed to the area in which these cells are perfectly intact and able to function. Consequently we expect an audiogram with a hearing loss curve showing a very faint slope.

So it is not very probable that a bass deafness in man with a steep hearing loss curve (cf. the continuous audiograms of our patients) is ever primarily based on an isolated loss of hair cells in the apical coils. It is possible that, in view of the anatomical relations, a disturbance in the blood circulation causes a degeneration of a certain part of the spiral ganglion.

As to the factors which might exercise an influence on the possibility of vibration of the basilar membrane, we may mention the changes in the pressure of the endolymphatic system, the "focal labyrinthitis" and the allergy of the internal ear. The loss of hearing in these cases is always explained by assuming that the basilar membrane can be less easily made to vibrate owing to a reduced elasticity or to an increase of mass by the precipitation of exudate.

Assuming that the hypotheses as to the reduction of elasticity and the increase of mass are valid, we must, in our opinion, also expect in these cases that the hearing loss curves in the audiogram have a very faint slope. This was actually found by SHAMBAUGH and TONNDORF.

The fact must be emphasized that in these cases an impairment of the organ of Corti is out of the question and that the hair cells are perfectly intact and go on functioning. The process is completely reversible in many cases; after a shorter or a longer time the hearing of the patients may become normal again. Here it is purely and simply a question of disturbance of intra-cochlear transmission, which appears in the whole range of frequencies, more pronounced in the bass region than in the region of the middle frequencies and here again stronger than in the region of the high tones. Therefore it is not justified to speak

of bass deafness in those cases in which a loss of hearing is found in the audiogram for all the usual frequencies (see introduction).

Arriving at a conclusion we may say that, an extensive mechanical damage of human hair cells in the apical coils of the cochlea being of rare occurrence, a bass deafness with such a steep hearing loss curve as found by us, is not based on a cochlear pathology.

Chapter III

SUPPLEMENTARY PATHOLOGICO-ANATOMICAL OBSERVATIONS

Since many authors (Shambaugh, Wright, Lindsay, Williams, Kos, Walsh, Hallberg, Opheim and Flottorp) are of the opinion that perceptive bass deafness is more or less typical for Ménière's disease (hydrops labyrinthi), at least in onsetting cases, it is important to look at the audiograms of patients suffering from Ménière's disease, whose temporal bones could be investigated after death and to note the alterations found in these cases.

On the other hand we have to check the alterations found in the temporal bones of patients, who had a hearing loss for the low tones without suffering from Ménière's disease.

Since 1938 several authors have published their histopathological findings in cases of Ménière's syndrome (Hallpike and Cairns 1938; Yamakawa 1938; Rollin 1940; Hallpike and Wright 1940; Lindsay 1942, 1944, 1946; Altmann and Fowler 1943; Cawthorne 1947; Arnvig 1947; Brunner 1948; Lempert, Wolff, Rambo, Wever and Lawrence 1952; Seymour 1954).

Over 20 cases are thus described in literature; 8 times an audiogram was made before the death of the patient. These 8 cases will be briefly surveyed.

Hallpike and Cairns (1938) described the case of a 28-year-old man who had been suffering from attacks of vertigo for 4 years. The hearing of his left ear was affected and was gradually getting worse; there was also a noise in his left ear. The audiogram was made 3 days before his death (figure 18); the patient died after an operation in which the left vestibular nerve was divided in the cerebello-pontine angle. In the left temporal bone the following pathological changes

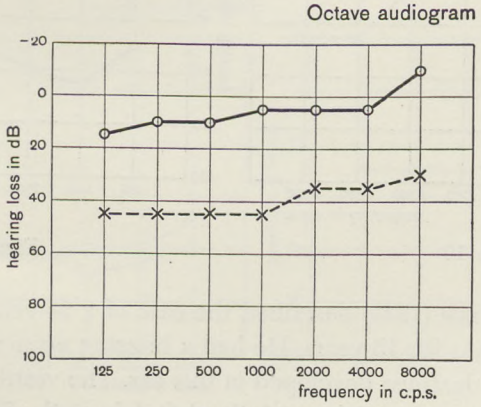


Figure 18

were found: gross dilatation of the saccule and scala media with obliteration of the perilymph cistern and of the scala vestibuli.

Degeneration and rupture of the wall of the anterior vertical membranous canal, together with the presence within its lumen of dense albuminoid coagulum. Degeneration of Corti's organ. Degeneration of the epithelium of the maculae and of the cristae of the semi-circular canals. Degeneration of the stria vascularis. Absence of the "normal" area of perisaccular connective tissue around the saccus endolymphaticus. The right temporal bone was not explored.

HALLPIKE and WRIGHT (1940) informed us of the case of a 29-year-old man who had had some defect in the hearing, tinnitus and a sensation of fullness in the left ear for 1 year. In addition there were attacks of vertigo. The audiogram was made 1½ months before his death (figure 19); the patient died as a result of repeated hemorrhages after intra-nasal intervention. In the left temporal bone there was a considerable distension of the scala media, which was present in all parts of the cochlea but which was particularly well marked in the posterior basal and anterior middle whorls where Reissner's membrane was thrown back into contact with the wall of the scala vestibuli over approximately one-third of its length. There was no change in the density of the spiral ganglion. There were no abnormalities of size or structure of the fibres of the VIIIth nerve. The organ of Corti showed considerable post-mortem degeneration, but the cells were otherwise normal. The stria vascularis was normal apart from severe post-mortem degeneration. The saccus endolymphaticus was for the greater part collapsed and the normal perisaccular connective tissue was absent. The right temporal bone showed the same post-mortem degeneration but was otherwise normal, except for an unusual degree of collapse of the saccus endolymphaticus.

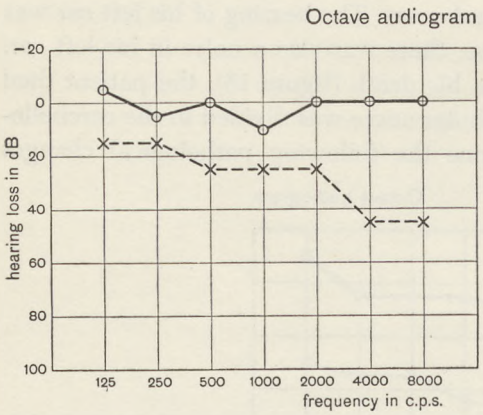


Figure 19

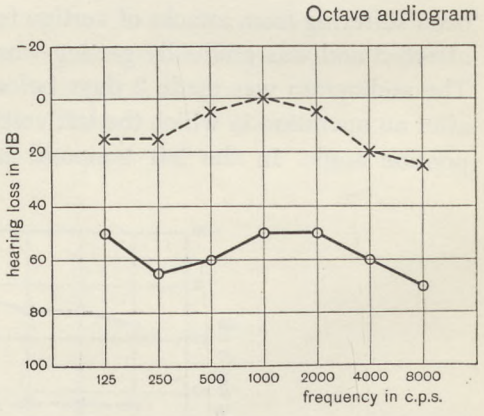


Figure 20

ALTMANN and FOWLER (1943) described the case of a 54-year-old man who had had attacks of dizziness for 16 years. He had a buzzing noise in the right ear and a steadily increasing deafness developed in this ear. The vestibular tests disclosed no differences between the right and the left labyrinth. The audiogram was

made 1½ months before his death (figure 20); the patient died after sectioning of the right auditory nerve. In the right temporal bone an extreme dilatation of the cochlear duct was found with herniation through the helicotrema; very marked dilatation of the ductus reuniens, the saccule, the utricle, the utricular and saccular ducts; herniation of the dilated utricle into the perilymphatic space of the proximal parts of the semicircular canals, slight dilatation of the proximal part of the endolymphatic duct. The dilated saccule covered the footplate of the stapes. The "perisaccular" tissue was absent. There were marked post-mortem changes in the end-organs and diminution in the number of ganglion cells and nerve fibres, particularly in the basal turn of the cochlea. In the left temporal bone there was no dilatation of the endolymphatic system. Post-mortem changes in the epithelium of the end-organs existed to the same extent as on the other side. Perisaccular fibrosis was somewhat less marked.

The same authors reported the case of a 52-year-old man who had had attacks of dizziness for 2½ years. He was becoming progressively deaf in the left ear; tinnitus was never noticed. The audiogram was made 1 week before his death (figure 21); the patient died of multiple myeloma. In the right temporal bone the cochlear duct showed extreme dilatation with herniation through the helicotrema, very marked dilatation of the proximal half of the ductus reuniens, and extreme dilatation of the saccule which covered the footplate of the stapes and bulged into the inner extremity of the external semicircular canal. The width of the utricle and the membranous semicircular canals was normal. There was slight dilatation of the saccular duct and of the proximal sinus of the endolymphatic duct. The perisaccular tissue was absent but there was an accumulation of pigmented cells in the subepithelial layers. There was a slight diminution in the number of ganglion cells and nerve fibres in the lower half of the basal turn of the cochlea. The aquaeductus cochleae was closed in its inner two thirds by loose reticular tissue which contained concretions. In the left temporal bone the changes were the same as in the right temporal bone. There were no pigmented cells in the fibrotic perisaccular tissue.

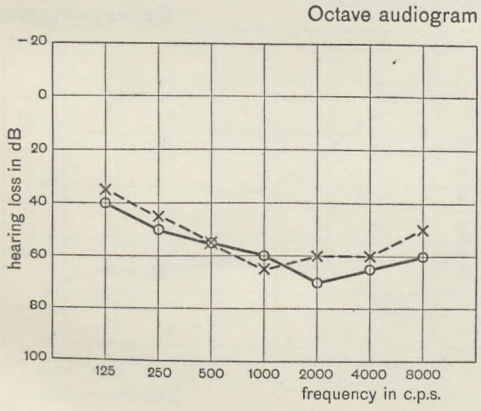


Figure 21

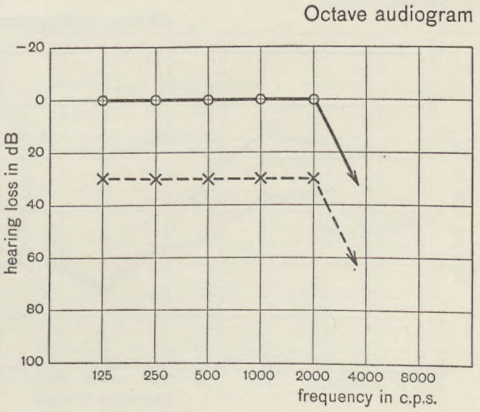


Figure 22

Lindsay (1944, 1946) described the case of a 47-year-old man who fell during an attack of vertigo, received a fracture of the skull and died from a subdural hematoma. He had been examined 3 years before and the diagnosis had been Ménière's disease. The right ear was normal for low tones up to and including 2048 cycles. An abrupt threshold loss was noted for frequencies above 2048 and a marked loss for all higher tones. The left ear revealed a loss of 30 dB for low tones, including 2048 cycles, and a rather sharp loss for tones above 2048 cycles (figure 22). A cold caloric test showed a diminished response on the left side. In the left temporal bone dilatation of the endolymphatic spaces was found with resulting distortion of membranes. The organ of Corti was present throughout and the hair cells seemed to be well preserved except for early post-mortem degeneration. The spiral ganglion appeared to have a normal content of nerve cells in the upper coils, but in the basal coil the cells were reduced in number. There was a generalized vascular congestion and extravasation of red blood cells. In the right temporal bone the pathological changes were limited to vascular congestion and hemorrhage in the modiolus and to degenerative changes in the spiral ganglion. The vascular congestion and hemorrhage in both temporal bones are explained by the terminal cranial injury, while the changes in the ganglion are probably directly connected with the hearing loss for tones above the frequency of 2048 cycles.

From the same author is the case of a 67-year-old man who had been suffering from an increasing deafness for 8 years, especially in his right ear. In addition there were recurrent attacks of vertigo. An audiogram was made 3 years before his death (figure 23). There was diplacusis, i.e. the frequencies 512, 1024 and 2048 had a lower pitch in the right ear. With the loudness balance test the difference in intensity was maintained, i.e. there was no recruitment. Resection of carcinoma of the cecum was followed by death from sudden cardiac failure. In the right temporal bone a marked hydrops was found involving the cochlear duct. Rupture of the saccule. Marked reduction of ganglion cells and nerve fibres in the basal coil. Early degenerative changes in Corti's organ. Hair cells

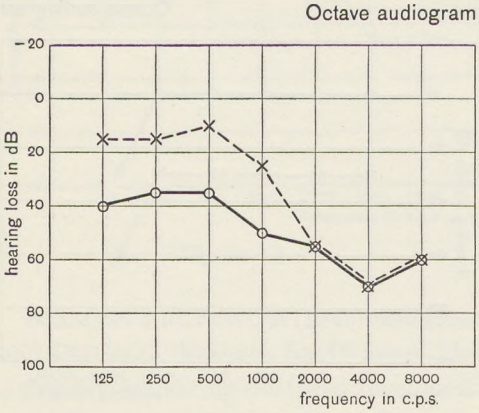


Figure 23

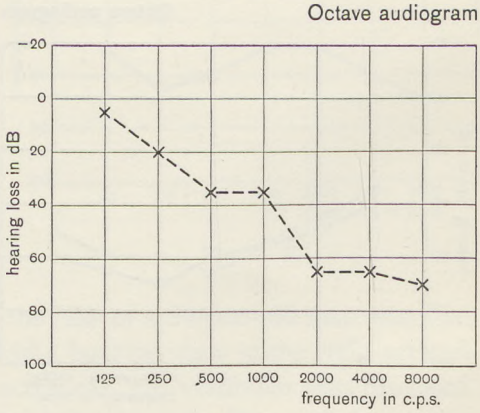


Figure 24

were probably reduced in number in the basal coil but showed early post-mortem degeneration throughout. The left temporal bone showed marked diminution of ganglion cells and nerve fibres in the basal coil of the cochlea. Degenerative changes in Corti's organ. The hair cells showed early post-mortem degeneration but seemed to show a relative reduction in number in the basal coil.

The third case described by Lindsay was a 55-year-old man who gave a history of ringing noises and deafness in the right ear for over 5 years. No mention of vertigo was included in the history. The audiogram was made 1 week before his death (figure 24); death occurred 2 days after laparotomy for suspected acute appendicitis. The patient was unable to hear loud shouts at the ear, when the left ear was masked by a Bárány noise apparatus. In the right temporal bone the cochlear duct was extremely dilated, obliterating the vestibular scala throughout much of its extent. Reissner's membrane was expanded to such a degree in the vestibule as to fill out the perilymphatic cistern and herniated for some distance into the small end of the horizontal canal and the common crus. The ganglion cells and nerve fibres were reduced to less than one-third of the normal number in the basal coil. The hair cells of Corti's organ showed early degenerative changes of the post-mortem type. The saccule was for the greater part collapsed. In the left temporal bone the structures of the inner ear were roughly normal. The ganglion cells and nerve fibres in the basal coil of the cochlea were reduced in number to less than one-third of normal. The hair cells in Corti's organ showed moderate post-mortem degenerative changes. There was also evidence of a reduction in their number in the basal coil.

Cawthorne (1947) reported the case of a 46-year-old man who had suffered from sudden bouts of giddiness for 4 years. Some time after the onset of these bouts he noticed deafness in the right ear. On caloric stimulation there was a deficiency of the right responses as compared with the left. The audiogram was made 9 months before his death (figure 25); the patient died from an acute lymphatic leukemia. In the right temporal bone the essential features observed

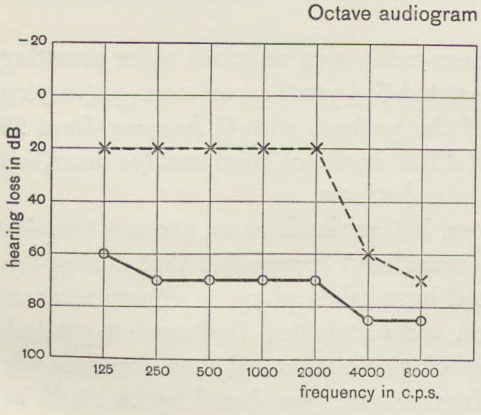


Figure 25

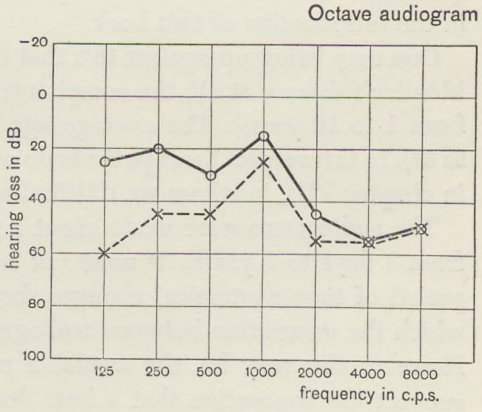


Figure 26

in previous cases, namely distension of the cochlear canal and changes in the perisaccal connective tissue, could be seen.

ODA (1938) published some observations on the pathology of impaired hearing for low tones. His material consisted of 35 ears from 19 individuals between the ages of 21 and 74. There was only one ear in which the hearing loss for the low tones was greater than that for the middle frequencies (figure 26). In the Weber's test the sound of the tuning fork was lateralized to the right ear. Otoscopical examination revealed no abnormalities. There were no complaints of tinnitus. The patient, a 47-year-old man, died from a glioma of the IIIrd ventricle. Histological examination showed a small otosclerotic focus in the central part of the stapes of the right ear; the annular ligament was not involved. There were no cochlear lesions more severe than in the control group with good hearing, no lesions that explained even the impairment for the high tones. In the left ear a histological otosclerosis was found, limited to the footplate of the stapes. Near the centre of the inferior margin, the articular cartilage and the annular ligament were replaced by the otosclerotic formation, so that here the footplate was in direct contact with the margin of the oval window but was not ankylosed to it. The cochlear lesions were within the limits of ears with good hearing. The author was inclined to ascribe the bass deafness in the left ear to the middle ear lesions found on histological examination, in spite of lateralization to the right in the Weber's test. The central nervous system was not histologically investigated, at least no data were furnished on this point.

DISCUSSION

It strikes us how regularly dilatation of the endolymphatic system was found in the mastoids of patients who had suffered from Ménière's disease. In the audiograms bass deafness does not come into prominence at all. It is even doubtful whether the loss of hearing for the low tones in the cases described by LINDSAY may be called bass deafness. For in this respect the curves of the audiograms do not come up to the requirements enumerated in the definition given in the introduction of this book.

One may bring up against this that these cases were no cases of an onsetting Ménière's disease at all; the complaints existed 5½ years on an average (varying from 1 to 16 years). The average age of the patients was 47 (varying from 28 to 67). In this respect these patients do not differ, however, from ours (as described in chapter VII), in whom we did find a bass deafness.

The audiograms were made about 1 year before death on an average (varying from 3 days to 3 years). It must not be considered improbable that during this period of time anatomical changes should have taken place, in consequence of which the correlation between audiogram and histological findings has got lost. However this may be, the available pathologico-anatomical data insufficiently support the conception that a bass deafness of the type found by us could be

based on a dilatation of the endolymphatic system, in this case a hydrops labyrinthi.

The case of bass deafness described by ODA, was thought to be based on an otosclerotic focus in the footplate of the stapes. Weber's test, however, showed that the tone was lateralized to the normal ear. No abnormalities were found in the cochlea; even the loss of hearing for the high tones could not be explained.

In this case we must take full account of the possibility that an affection of the central auditory pathways must be held responsible for the loss of hearing for the high tones on both sides and the loss of hearing in the bass region on one side.

Chapter IV

BASS DEAFNESS DUE TO LESIONS IN THE AUDITORY PATHWAYS

Neff (1947) measured the hearing of cats by means of the conditioned response method before and after partial section of the eighth nerve at the entrance of the internal auditory canal on one side, the cochlea of the opposite side having been destroyed prior to the initial hearing tests. As signs of vestibular disturbance were often present, we must suppose that the vestibular portion of the eighth nerve was severed together with the cochlear portion.

Stimulus thresholds were determined for the frequencies 125, 250, 500, 1000, 2000, 4000 and 8000 c.p.s. and in some cases for the frequencies 60 and 15.000 c.p.s. No hearing losses were found when the lesions, as estimated at the time of operation, involved one-tenth to one-half of the nerve. Lesions involving approximately one-half or more of the nerve, produced hearing losses for frequencies of 1000 c.p.s. and above. No significant hearing losses were found for frequencies below 1000 c.p.s. If loss of hearing occurred, it included all frequencies higher in the scala than the lowest for which there was any loss. For example, if a loss occurred at 4000 c.p.s., an equal or greater loss occurred at 8000 and 15000 c.p.s. When the lesions were very large or nearly total the resulting hearing loss was complete at all frequencies.

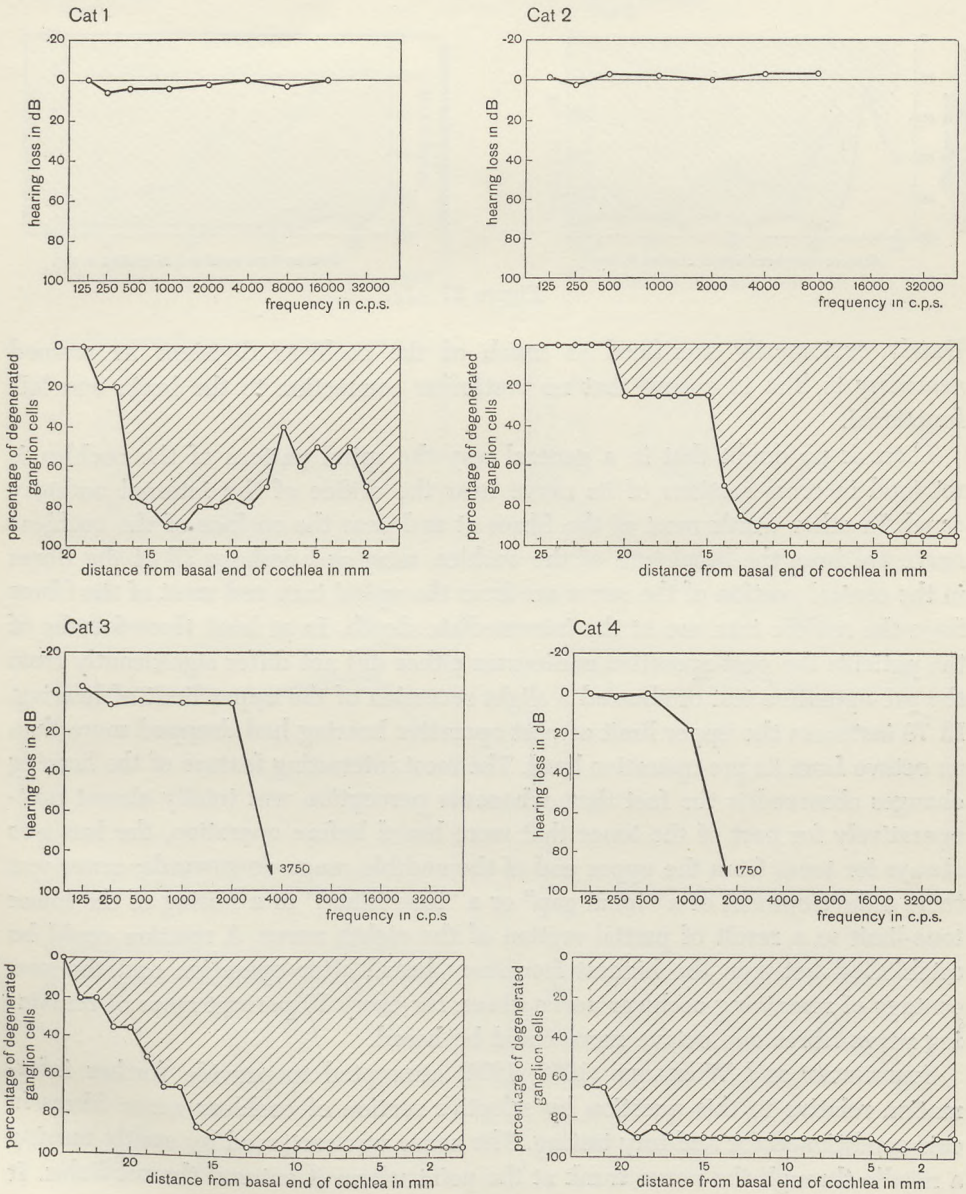
Wever and Neff (1947) presented the results of cochlear response tests and histological examination of the cochleas of the cats described above. The cochlear responses recorded were for all animals but one within the range usually found for normal animals. This one showed normal sensitivity, in the cochlear response tests, for tones below 700 c.p.s., a slightly reduced sensitivity from 700 to 3000 c.p.s., a severe reduction from 3000 to 7000 c.p.s. and complete absence of response at 8000 c.p.s. and above.

Histological examination of the cochleas revealed partial degeneration of neural elements but normal hair cells in all animals except the animal mentioned; in this animal the hair cells in the basal part of the cochlea were atrophied. A period of at least eleven weeks elapsed between the time of sectioning the eighth nerve and the cochlear response tests.

In figure 27 the audiograms of 6 cats have been brought together, made according to the conditioned response method. On histological examination, it proved that the hair cells of the organs of Corti of these animals were always unimpaired. Under each audiogram a diagram is found which renders the percentage of ganglion cells that proved to be degenerated at various distances from the basal end of the cochlea when histologically examined. In this way we can

see immediately whether there is any correlation between the audiogram and the histological findings or not.

GUILD (1953) presented the results of hearing tests before and after section of the eighth cranial nerve in patients with Ménière's disease; the operations were performed by Dandy. The operative procedure varied only in the amount of the eighth nerve that was divided; the surgeon wished to sever all of the vestibular division and, if possible, to preserve all of the cochlear division of the nerve. When, in a patient, the boundary between the two divisions was not distinct,



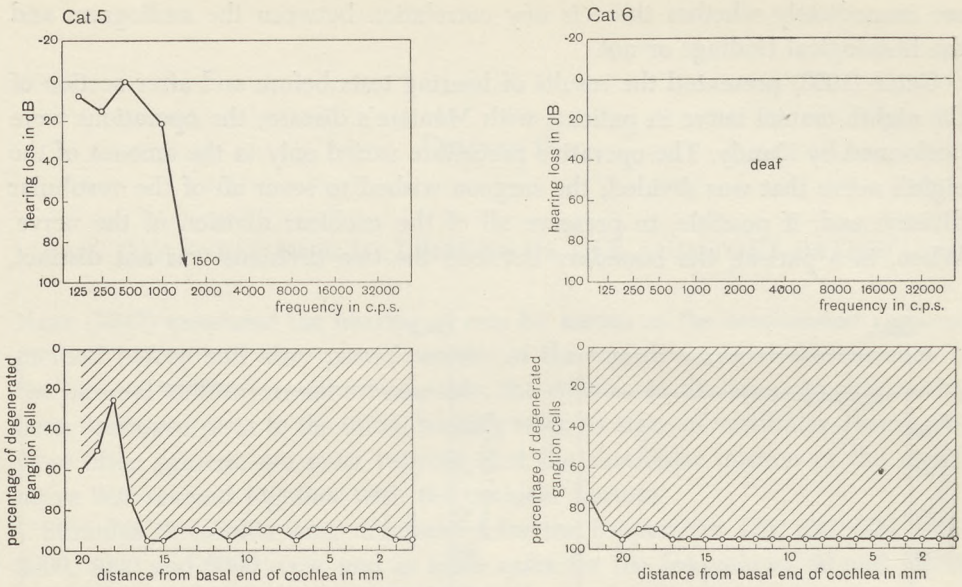


Figure 27

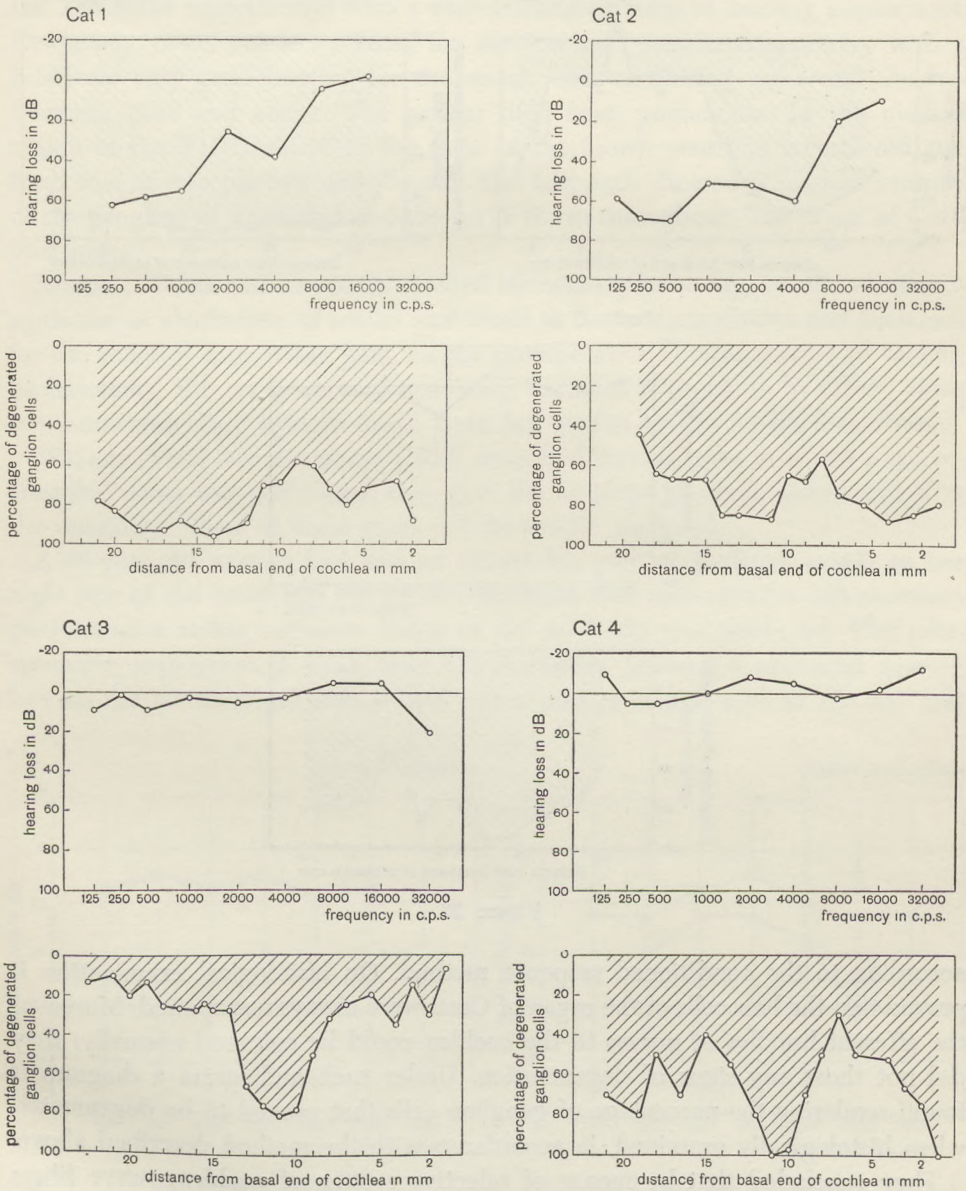
Dandy customarily sacrificed as much of the cochlear divisions as seemed necessary to him to ensure that no vestibular connection to the brain was left functional.

There is no doubt that in a general way the spiral pattern of the cochlea is retained in cross sections of its nerve near the orifice of the internal auditory canal. In other words most of the fibres at and near the surface of the cochlear nerve are from the basal turn of the cochlea, most and perhaps all of the fibres in the central portion of the nerve are from the apical turn and most of the fibres from the middle turn are at the intermediate depth. In at least three-fourths of the patients the post-operative audiogram either did not differ significantly from the pre-operative test or showed a slight recession of the upper limit of hearing. In 78 instances the upper limit of post-operative hearing had dropped more than an octave from its pre-operative level. The most interesting feature of the hearing changes observed is the fact that, whenever perception was totally absent post-operatively for part of the tones that were heard before operation, the loss was always for tones from the upper end of the audible range downwards; never was there a development of a "tonal gap" or a "tone island" or a raising of the lower tone-limit as a result of partial section of the eighth nerve. A relation could be established between the highest frequency heard after operation and the percentage of remaining cochlear nerve fibres: for every 10 percent more of remaining nerve fibres one octave more could be heard.

Schuknecht and Woellner (1953, 1955) attempted to provide further information relative to this question by selective section of cochlear nerve fibres in cats conditioned for auditory testing. The nerve was sectioned by gently moving a needle through the nerve trunk at the point where it entered the modiolus. It

was thought that this technique would sever the fibres supplying the apical and middle turns and the upper part of the basal turn while leaving the fibres to the lower basal turn intact. The histological findings show that, in reality, this was only partially accomplished. One week after the operation, hearing tests were made. The auditory thresholds showed no significant changes in the interval between the first tests made one week post-operatively and the last tests performed about four weeks post-operatively.

In figure 28 the audiograms of 7 cats have been brought together, made



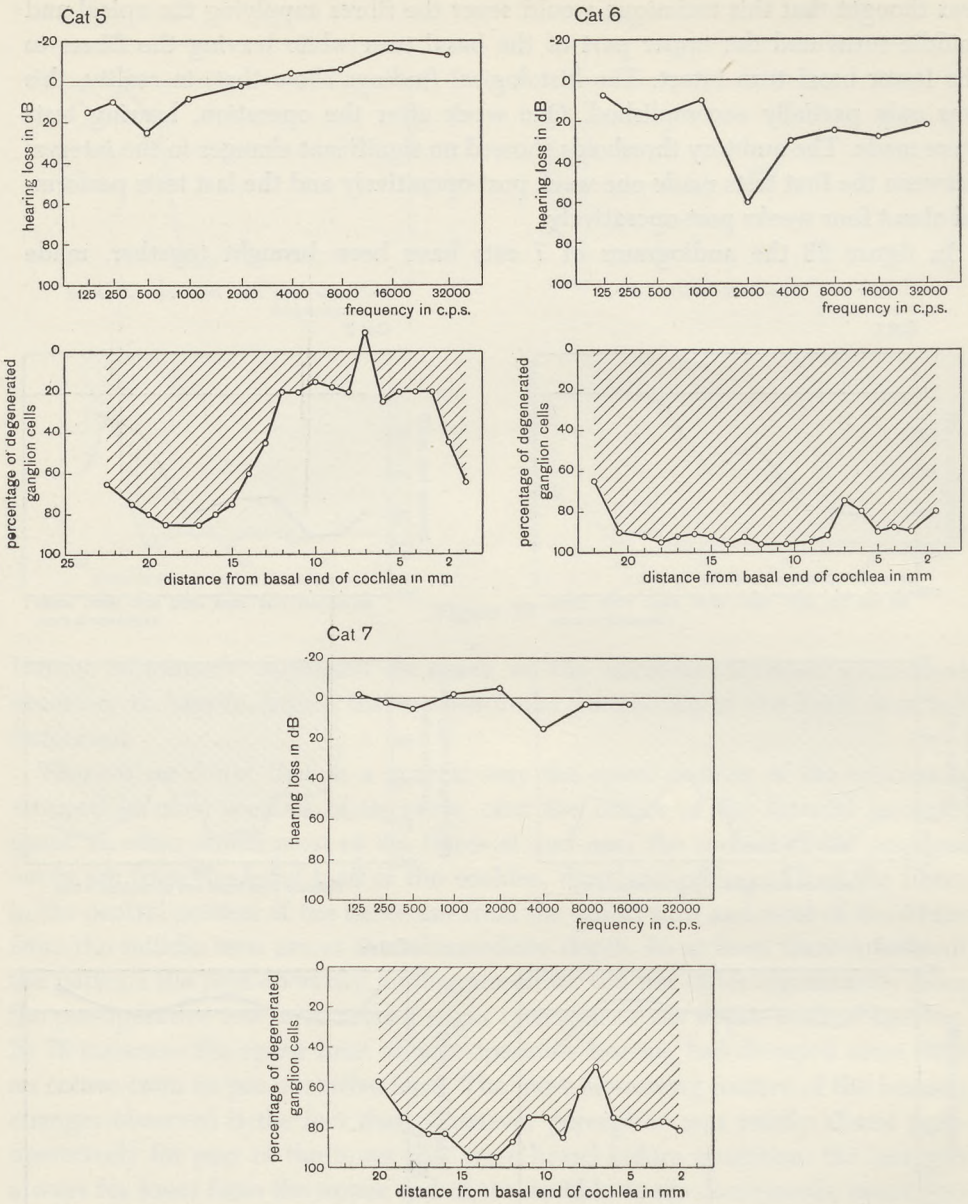


Figure 28

according to the conditioned response method. On histological examination it proved that the hair cells of the organ of Corti were always unimpaired. Moreover the efferent bundles of nerves to the cochlea could be coloured normally; they did not show any signs of degeneration. Under each audiogram a diagram is found rendering the percentage of ganglion cells that proved to be degenerated when histologically examined, in accordance with the method described above.

They concluded that by means of selective section of cochlear nerve fibres

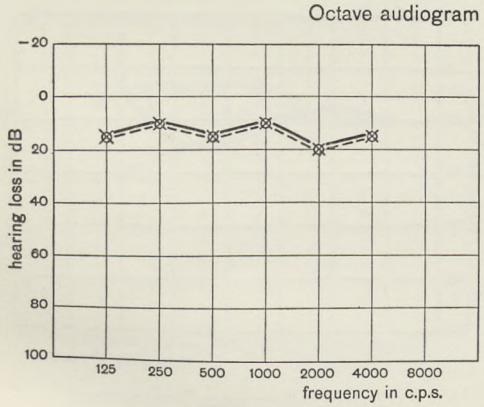
restricted hearing losses could be produced for both low tones and high tones and they thought that these findings supported the conception that there is spatial representation in the spiral ganglion for low tones as well as high tones. Their experiments indicate that the threshold for any frequency of the auditory spectrum remains normal when 20 to 40 per cent of the spiral ganglion cells remain in the region of the cochlea serving that frequency, but that a threshold elevation is likely to exist when less than 20 per cent of the ganglion cells remain.

Krassnig (1924) described the case of a patient suffering from a bulbar symptom complex (symptoms showing an affection of the VIIth, the XIIth and the Xth nerve respectively) with a very interesting form of hearing impairment. Essentially there was a deafness for the low and middle frequencies with a relatively very good hearing for the watch, the whispering voice and the frequencies 2048 and above. The patient died from pneumonia. In the nuclear region of the VIIIth nerve in the floor of the fourth ventricle several old and fresh foci of encephalitis were found. The labyrinth showed a marked atrophy of the ganglion of Rosenthal and the nerve fibres throughout. The organ of Corti was intact.

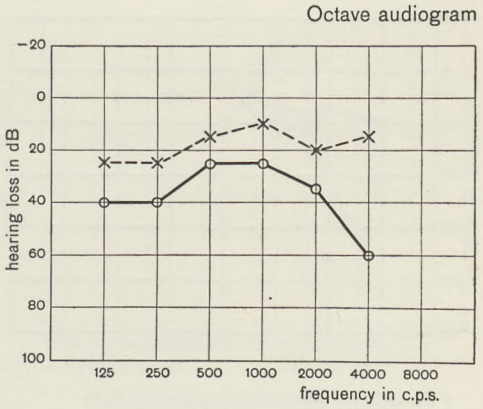
Spiegel, Wycis and associates devised stereoencephalotomy, i.e. the electrocoagulation or electrolysis of nuclei and tracts in the mesencephalon and thalamus for the relief of intractable pain. In the postoperative clinical picture of mesencephalotomy, the occurrence of a hearing defect is a probable by-effect, since the pain-conducting spinothalamic tract lies medial to the medial lemniscus.

Walker (1942) and Saltzman (1952) described several cases in which a stereoencephalotomy was performed; they gave the audiograms made before and after the operation. Some of these cases will be briefly surveyed.

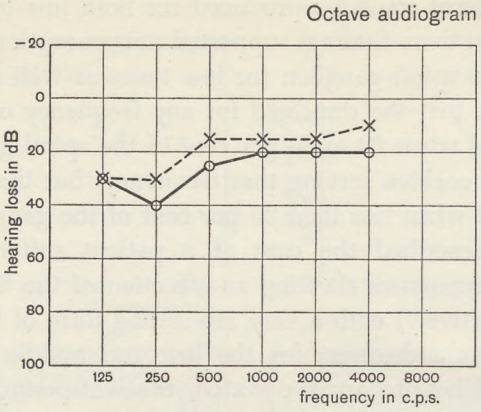
A 62-year-old woman had had an intractable pain of thalamic origin on the right side of the body and the right extremities. She underwent a left mesencephalotomy; a rather extensive lesion of the midbrain was produced. The postoperative audiogram, 1 week after the operation, showed a profound hearing loss on the contralateral side. There was a greater impairment of the low and



before operation



1 week after operation

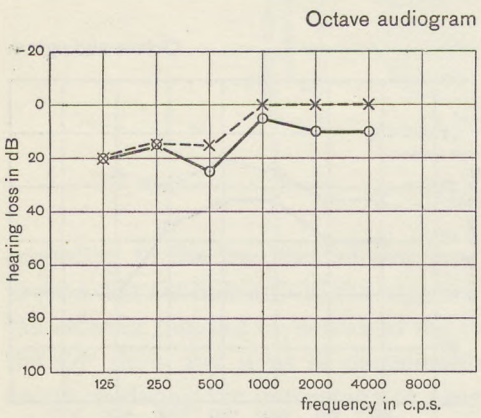


4½ months after operation

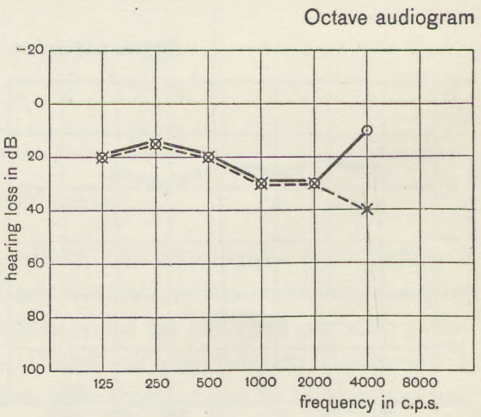
Figure 29

high tones than of the middle frequencies. On the ipsilateral side a slight impairment of the low tones was shown. The audiogram made four months later showed phenomenal recovery, but a bilateral defect for the low tones was present (figure 29).

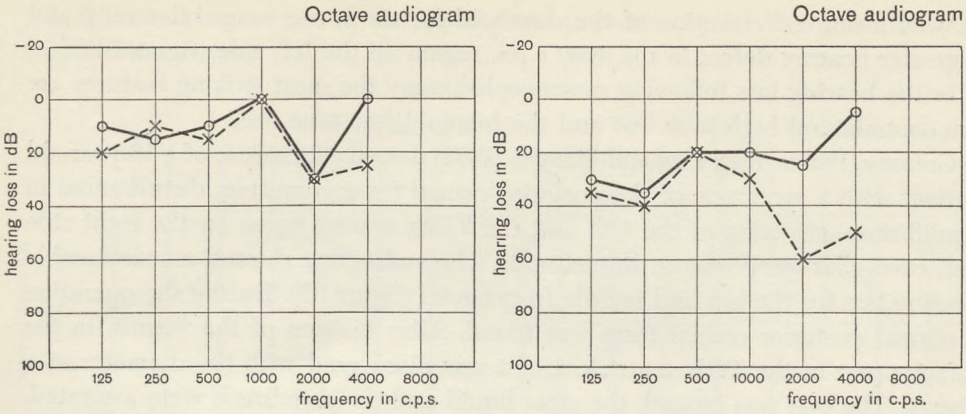
A 29-year-old woman was admitted with the complaint of intractable "sciatic pain" on the right side. No relief was obtained from conservative treatment. A left mesencephalotomy was performed. The audiogram, 8 months after the operation, showed a bilateral defect for the low tones and a contralateral impairment for the middle and high tones. The patient was known to have had normal hearing prior to this operation. The patient returned with an intractable pain in both lower extremities. A bilateral mesencephalotomy was performed. The audiograms, 2 weeks after the second operation, showed bilateral impairment for the middle and high tones. Some 4 months later there was a complete recovery in the 1000 c.p.s. region. The patient came back because of dysesthesia to pain,



8 months after first operation; left mesencephalotomy



2 weeks after second operation; bilateral mesencephalotomy



4 months after 2nd operation; bilateral mesencephalotomy

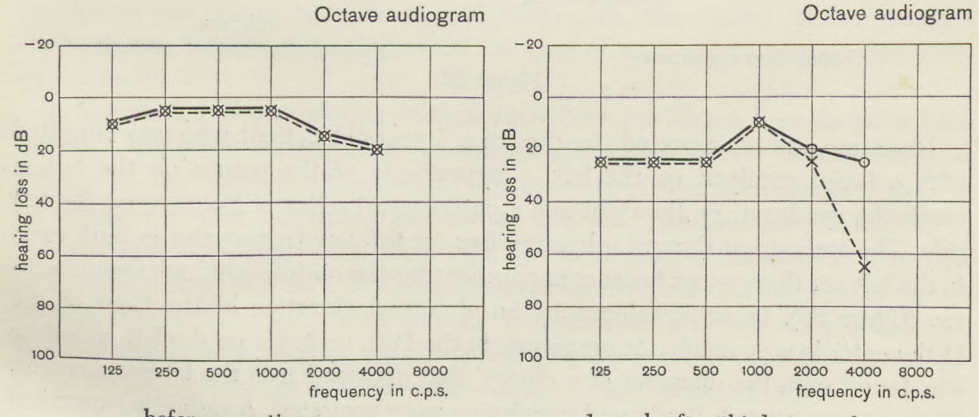
11 months after 3rd operation; right mesencephalotomy

Figure 30

touch and temperature. A right mesencephalotomy was performed. The audiogram, 11 months after the third operation, showed a contraction of the auditory field which was most pronounced in the left ear, a bilateral rise in the threshold for the lower tones, and a marked contralateral loss for high tones that followed the third operation, a right mesencephalotomy (figure 30).

Tinnitus appeared soon after the first operation but after the third operation it became more pronounced on the left side. Speech sounded shrill and was unintelligible in the left ear.

A 52-year-old man was admitted with the diagnosis of gastric crises. His complaint was an unbearable pain in the stomach for the last ten years. The mesencephalotomy was performed in several stages. Small lesions were placed in the midbrain on the right side; some three weeks later these lesions were enlarged. After another three weeks large lesions were placed in the left mesencephalon. The audiogram made one week after the third stage of the operation



before operation

1 week after third stage of operation

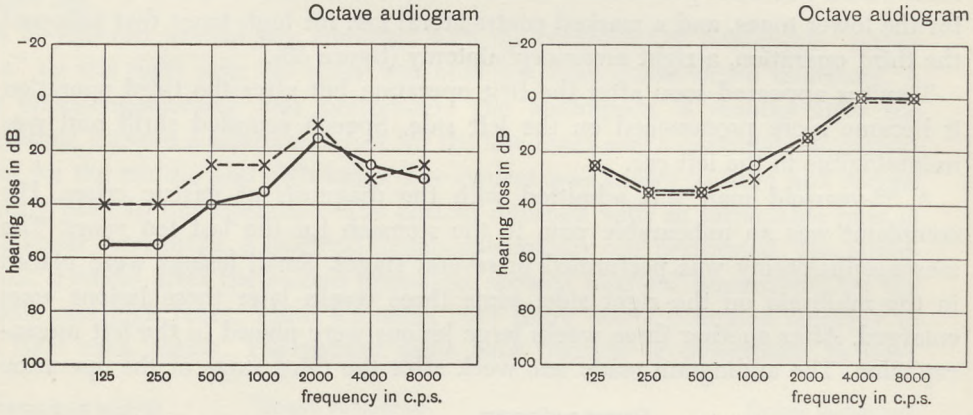
Figure 31

showed a marked elevation of the threshold for the lower tones bilaterally and a greater hearing defect in the 4000 c.p.s. region on the left side (figure 31).

In the hearing loss following mesencephalotomy the most striking features are the contralateral high tone loss and the bilateral low tone loss.

GREINER, PHILIPPIDÈS, ISCH and MENGUS (1953) described the case of a 13-year-old patient with a syndrome of the posterior cranial fossa: vomiting, disturbances in equilibrium, affection of the Vth and the VIIth cranial nerve on the right side and cerebellar symptoms on the left side. The audiogram showed a considerable hearing loss for the low and middle frequencies (figure 32). During the operation a normal posterior cranial fossa was found. After incision of the vermis, in the inferior part of the IVth ventricle a cyst was discovered with the diameter of a cherry. The cyst was incised, the clear liquid and the membrane were aspirated. No tumour was found.

One year after the operation there was a great improvement; the patient did not complain of deafness. Four years after the intervention there were neurological signs of a relapse. The audiogram showed again a hearing loss for the low and the middle frequencies. Operation revealed the IVth ventricle to be enlarged and filled by several cysts.



before first operation

relapse;
1 year before second operation

Figure 32

The same authors reported the case of a 7-year-old patient who was admitted with a facial paralysis on the left, a hypesthesia of the cornea on the left, a cerebellar syndrome on the right and a pathological reflex of Babinski on the left side. The audiogram showed a hearing loss for the low frequencies in both ears; in the left ear there was a hearing impairment for the middle and high frequencies too (figure 33). In encephalography an abnormal elevation of the floor of the IVth ventricle was visible. At operation, in the IVth ventricle on the left, a nodule was found with the diameter of a cherry, that extended into the lateral recessus. The cyst was incised; liquid and membrane were aspirated. A radio-therapeutical aftertreatment was carried out. At control examination, 1½ years later, there

was a general improvement; the hearing had improved by 20 dB for the low and middle frequencies.

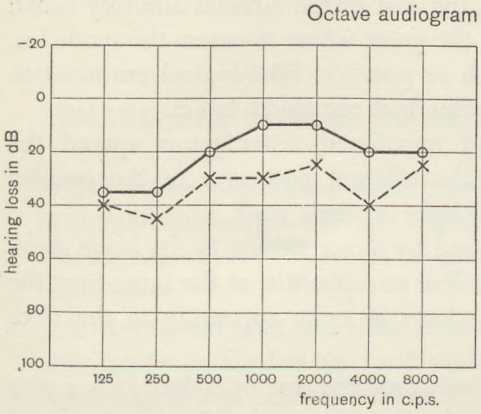


Figure 33

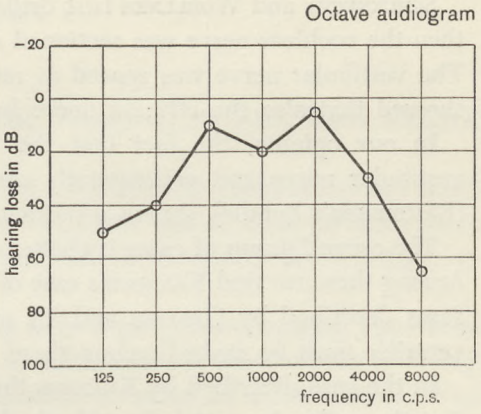


Figure 34

LUNDBORG (1955) described the case of a 58-year-old woman with an acoustic tumour on the right side, verified at operation. The audiogram showed nerve deafness for low tones and for the frequencies above 4000 c.p.s. on the right side; the left side was normal (figure 34).

DISCUSSION

In order to get a convenient arrangement of the various findings that have been described, it is advisable to group them according to the level at which the auditory pathway was impaired.

The first group of cases is characterized by a lesion of the VIIIth nerve. The consequence of this was that generally there was degeneration of ganglion cells while the organ of Corti remained intact. In this group we find the experiments by WEVER and NEFF, SCHUKNECHT and WOELLNER, the results of the Dandy's operations as described by GUILD and the case of an acoustic tumour described by LUNDBORG.

It is remarkable that neither WEVER and NEFF nor GUILD ever found a bass deafness following on a partial section of the VIIIth nerve. SCHUKNECHT and WOELLNER did find this while also LUNDBORG found a bass deafness in a case of an acoustic tumour disturbing the normal course of the cochlear fibres.

SCHUKNECHT and WOELLNER think that loss of hearing is caused for a certain frequency if less than 20 % of the ganglion cells are intact in the region of the cochlea where the frequency in question is perceived. This rule does not always apply to every case of their own material (c.f. their cats 1 and 2 with cat 5), while in cat 1 of WEVER and NEFF there was no bass deafness either.

Apart from this there is some difference in the techniques of these investigators. WEVER and NEFF sectioned the VIIIth nerve partially at the entrance of the

internal auditory canal; they sectioned the vestibular nerve and part of the cochlear nerve (Dandy's technique). The results obtained square with those obtained by GUILD.

SCHUKNECHT and WOELLNER first drilled the roof of the internal auditory canal; then the cochlear nerve was sectioned at the point where it enters the modiolus. The vestibular nerve was spared as much as possible. Histological examination showed that also the efferent nerve bundles had remained intact.

In our opinion, the fact that the last mentioned investigators spared the vestibular nerve and consequently also the efferent pathways to the cochlea (Rasmussen's bundle), throws a particular light on their work.

The second group of cases is characterized by abnormalities in the brain stem. Among these we find KRASSNIG's case of bulbar encephalitis; at the same time the cases described by GREINER and his associates of cysts appearing in the IVth ventricle must be ranked among these.

In the case described by KRASSNIG the bass deafness was onesided. It is a pity that the author has not indicated whether the foci of encephalitis were localized on the right or on the left side of the brain stem. Ganglion cells and nerve fibres were for the greater part degenerated. The organ of Corti was intact.

In the cases of GREINER and his associates the bass deafness existed on both sides and was of a varying nature. Nothing is known of the condition of the ganglion cells and the organ of Corti, as a histological examination of the cochlea could not take place. We are entitled to suppose that these structures had not been damaged in a serious way seeing that the loss of hearing was reversible.

The third group of cases is characterized by a lesion of the auditory pathway at an even higher level. We refer to the stereoencephalotomies, the effects of which on the hearing were described by WALKER and SALTZMAN.

After such a stereoencephalotomy, at least when the lateral lemniscus is sectioned, we generally see a contralateral loss of hearing for the high tones and a bilateral loss of hearing in the bass region.

The loss of hearing for the high tones is often reversible and is believed to be based on a temporarily reduced number of functioning nerve fibres as a result of postoperative edema.

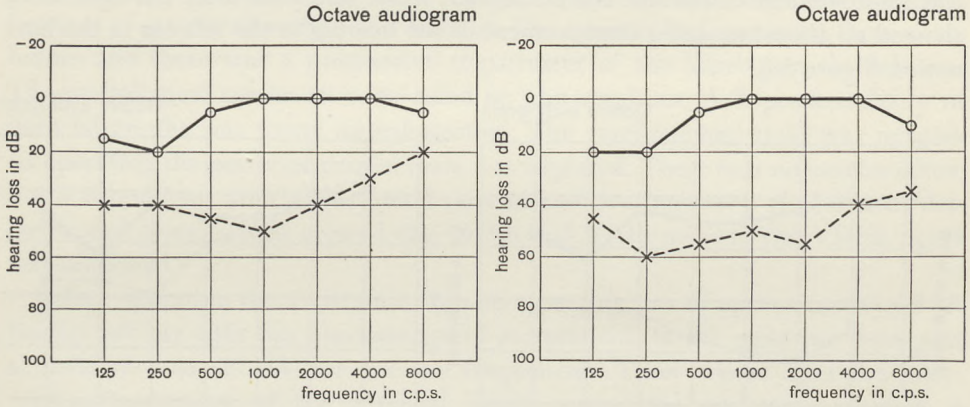
No explanation sofar has been found for the bilateral loss of hearing for the low tones which is mostly of a permanent nature.

Chapter V

REMARKABLE BEHAVIOUR OF THE HEARING IN THE BASS REGION

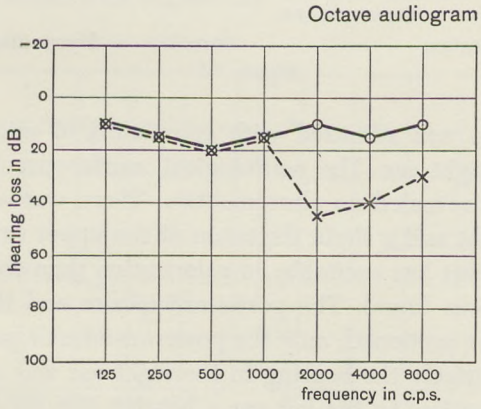
In 1937 Aubry and Ombrédanne published a report on the indications and results of intracranial surgery of the VIIIth nerve. Three out of their 46 cases are of special interest to us, because of the remarkable behaviour of the hearing in the bass region.

A 41-year-old man had been suffering from a progressive hearing impairment and tinnitus in the left ear for 3 years; in addition there were attacks of vertigo. Neurological and cardio-vascular examination revealed no abnormalities. While



before operation

2 months after operation

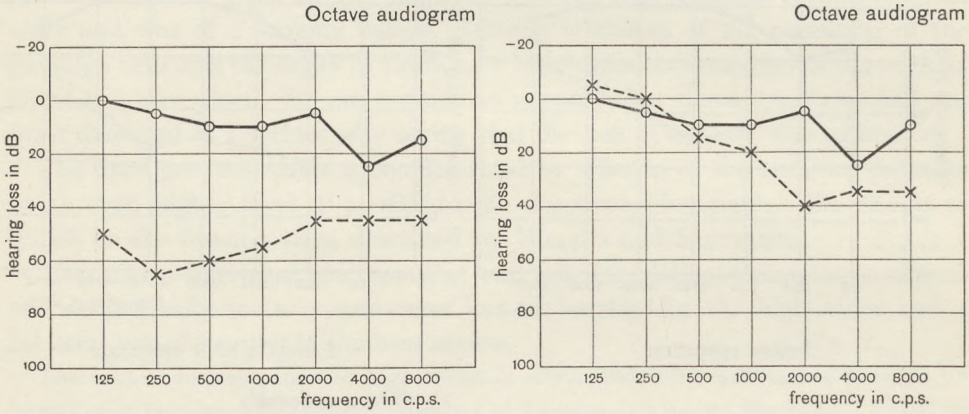


1½ years after operation

Figure 35

walking with closed eyes there was a slight deviation to the left. The left labyrinth was less excitable on calorization than the right. The patient was operated on with partial section of the VIIIth nerve. There was no apparent division of the nerve; the vestibular portion was cut. No arachnoiditis was found. Postoperatively a progressive improvement of the hearing was observed; 15 days after the operation the hearing was essentially the same as before the intervention, but an audiogram made 1½ years later, showed a remarkable improvement of the hearing in the left ear in the bass region (figure 35).

A 26-year-old man complained of deafness and tinnitus in the left ear, with attacks of vertigo and headache. Neurological and cardio-vascular examinations were completely negative. There was a slight leftsided adiadochokinesis and hypotonia of the left extremities. On calorization the left labyrinth was less excitable than the right. No arachnoiditis was found. The VIIIth nerve was thin and atrophic. The posterior fibres of the nerve were superficially sectioned. Already one day after the operation the hearing in the left ear had improved and this improvement continued. An audiogram made one year after the operation, showed an almost amazing improvement of the hearing in the left ear in the bass region (figure 36).



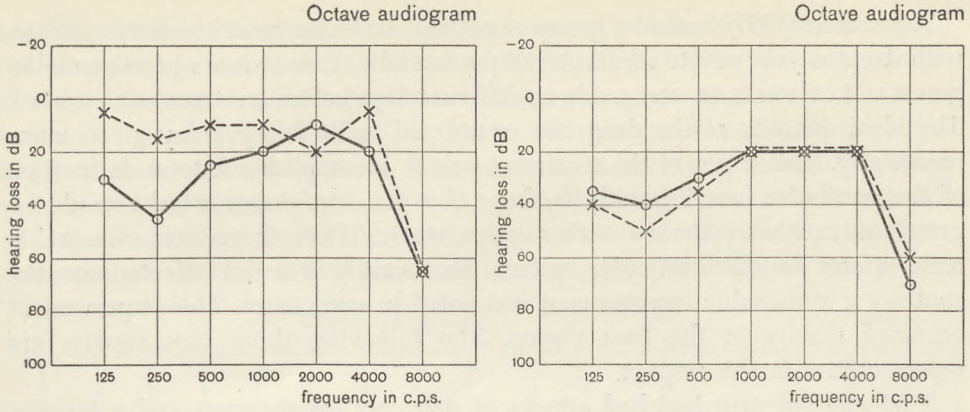
before operation

1 year after operation

Figure 36

A 50-year-old man was admitted with complaints of attacks of vertigo and hearing loss in the right ear. The neurological, cardio-vascular and ophthalmological examinations revealed no abnormalities. There was a slight spontaneous nystagmus to the right and a slight deviation of the upper extremities to the left. The right labyrinth was less excitable on calorization than the left.

No arachnoiditis was found. The posterior-superior and the anterior parts of the VIIIth nerve were sectioned; only the posterior-inferior part of the nerve was left intact. Postoperatively the hearing in the right ear was essentially the same as before the intervention. In the left ear a hearing loss for the low frequencies had occurred, but this bass deafness disappeared again in the months following the operation (figure 37).

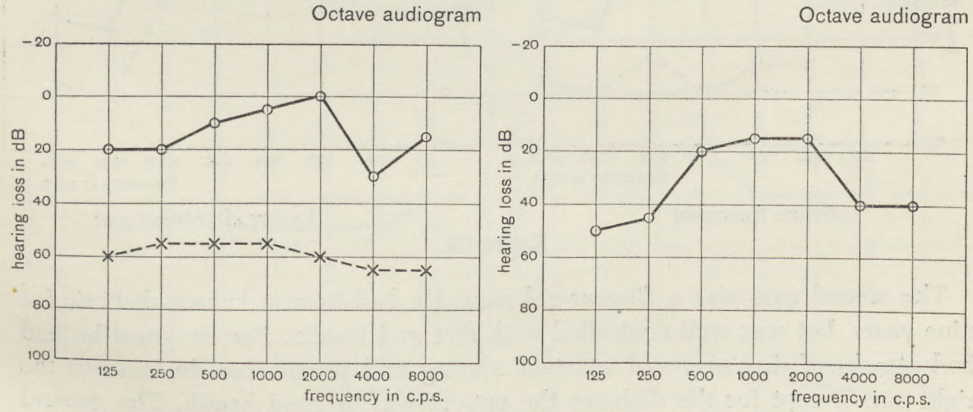


before operation 12 days after operation

Figure 37

From the same point of view a case reported by THIÉBAUT, ROHMER, GREINER and MENGUS is of interest to us. It was a 50-year-old man who was admitted because of attacks of vertigo and tinnitus. The tinnitus had appeared three years before and there was a progressive impairment of the hearing in the left ear. The neurological examination revealed no abnormalities. A hypo-excitability of both labyrinths was found on calorization. The cerebrospinal fluid was normal. At operation the posterior cranial fossa was explored. There was no arachnoiditis, but a voluminous arteria was found (arteria auditiva interna?), of abnormal diameter and place, which crossed the VIIIth and VIIth nerves. The VIIIth nerve was sectioned.

Before operation the audiogram showed a hearing loss of approximately 60 dB for the left ear with the phenomenon of recruitment; in the right ear there was a moderate loss (20 dB) for the low frequencies. Three weeks after the intervention (sectioning of the leftsided VIIIth nerve) the audiogram showed a distinct bass deafness in the right ear; apart from that there was some hearing loss for the higher frequencies (figure 38).

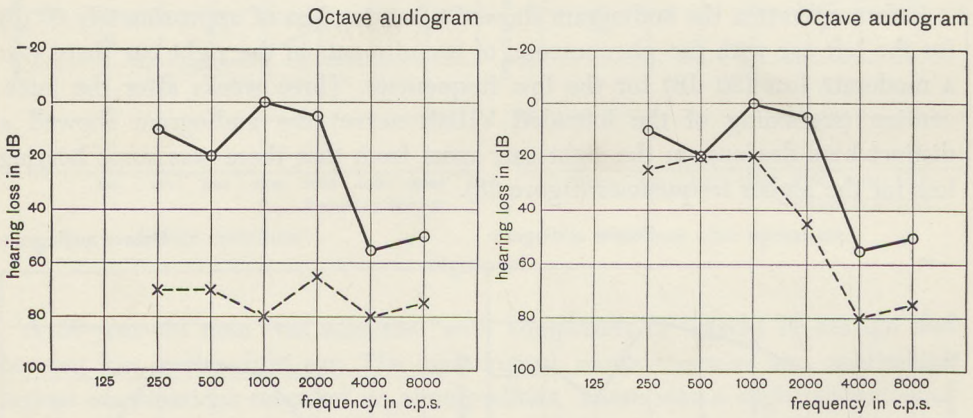


before operation 3 weeks after operation

Figure 38

SCHUKNECHT (1957) treated a group of patients suffering from Ménière's disease, with streptomycin sulfate administered parenterally. Two or more baseline audiograms and caloric tests were made on different days before treatment was started. The ideal quantity of the drug was considered to be 2 g per day given intramuscularly. The object of the treatment was to accomplish sub-total destruction of the vestibular function with the hope that the vertiginous attacks would be greatly attenuated without creating severe ataxia. While there were remarkable toxic effects for the vestibular system, the hearing was not affected; on the contrary a remarkable improvement was noted in some cases. This improvement occurred mainly in the bass region. The following three case reports are representative in this respect.

A 56-year-old man had had attacks of dizziness for six years and a hearing loss in the left ear. Audiometrical study revealed a 60–80 dB perceptive hearing loss in the left ear with complete loudness recruitment. The discrimination score was 8 % at 98 dB above the average normal speech threshold. The caloric tests gave equal responses in each ear but with decreased amplitude on the left. After parenteral streptomycin therapy, during which 50,5 g of the drug was given, there was no response to ice water caloric test in the left ear. Since the treatment started, the patient had still had attacks of fullness and buzzing in the left ear coming on at intervals from one to six months but without dizziness. Audiometrical studies revealed an over-all hearing improvement in the bass region since the treatment had started, with thresholds fluctuating from 20 to 30 dB in the diseased ear and coinciding with the patient's subjective impression of his auditory acuity (figure 39).



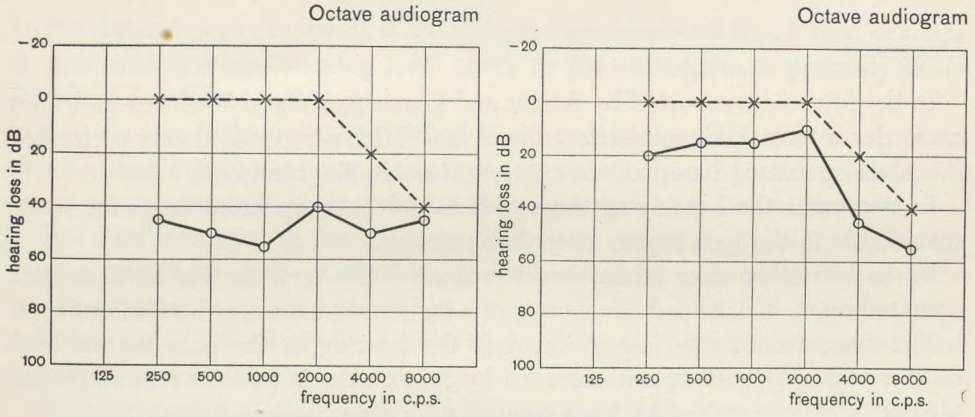
before treatment

4 years after treatment

Figure 39

The second case was a 49-year-old man. He had been a known diabetic for nine years, but was well controlled with diet and insulin. For six years he had had attacks of dizziness and constant roaring and progressive deafness in the right ear. Except for the diabetes the patient was in good health. The general examination revealed no abnormalities. The right labyrinth was less excitable

than the left on calorization. Audiometrical studies persistently revealed a perceptive type of pure tone threshold loss of 40–55 dB and a discrimination score of 62 % in the right ear. Streptomycin sulfate was given intramuscularly in a dosage of 1 g twice daily. When the treatment was discontinued, 89 g of the drug had been administered and there was only a very mild caloric response left in both ears. The roaring tinnitus disappeared during the treatment and did not recur. The auditory thresholds in the diseased ear were improved in the bass region immediately after treatment and remained improved (figure 40).

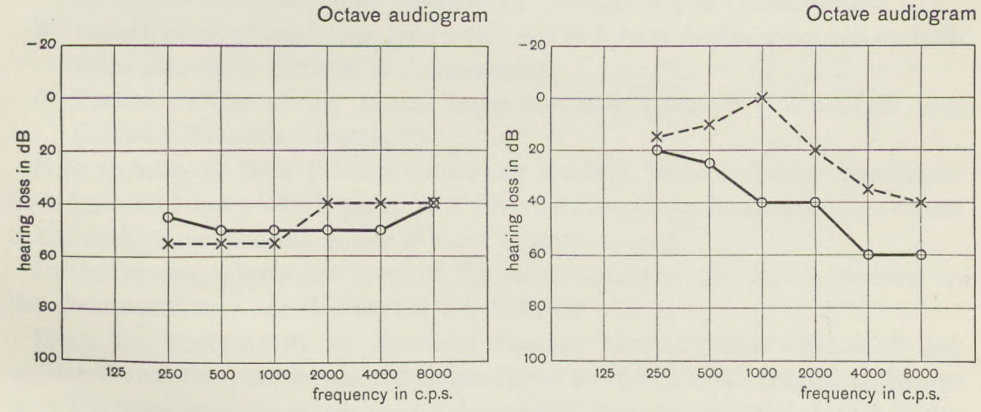


before treatment

33 months after treatment

Figure 40

The third patient was a 32-year-old man who had been having attacks of dizziness for five years; in addition there was a progressive loss of hearing in both ears. He had a constant, high pitched tinnitus, worse in his right ear. General examination revealed no abnormalities. Speech discrimination scores were 74 % on the right and 84 % on the left (both tested at an intensity of 88 dB, using 70 dB white noise masking in the opposite ear). The caloric responses of both labyrinths were of a reduced amplitude; the left labyrinth was less excitable



before treatment

34 months after treatment

Figure 41

than the right. Although there were certain features of migraine (blurring of vision and headache) and of epilepsy (sequela of drowsiness), the most prominent symptoms involved the auditory and vestibular systems. About three weeks after the beginning of the streptomycin therapy, the audiograms revealed an average gain of about 15 dB throughout the auditory spectrum. When last tested, thirty-four months after treatment, there was a mild caloric reaction to ice water in both ears and the auditory thresholds had remained at the improved level in the bass region (figure 41).

Discussion

In the four cases reported by Aubry and Ombrédanne and Thiébaut and their associates, a partial or complete section of the VIIIth nerve was always performed (Dandy's operation) based on an existing Ménière-like syndrome.

In two cases the hearing of the ear that had been operated upon improved remarkably in the bass region after the operation.

In the two other cases a bass deafness arose in the very ear that had not been operated upon. This bass deafness was of a temporary nature, at least in one case.

The same remarkable improvement of the hearing in the bass region, both on one and on two sides, was observed by Schuknecht in patients suffering from Ménière's disease, who had been treated with streptomycin.

Chapter VI

PRELIMINARY CONCLUSIONS

In the discussion of chapter II it has already been observed that a bass deafness in man with a steep "hearing loss" curve in the audiogram is probably never based on a cochlear pathology.

Nor do the pathologico-anatomical findings (chapter III) confirm the conception that a bass deafness of the type described by us, would generally be based on an affection of the peripheral sense organ.

The data, recorded in the chapters IV and V, very strongly suggest that the explanation of the origin of the bass deafness meant by us, must be found in the nervous system.

Before going into this in further detail, it will be useful to summarize the most important conclusions that have been reached in the preceding chapters:

1) After partial section of the VIIIth nerve, bass deafness can only arise if the efferent pathways to the cochlea (Rasmussen's olivocochlear bundle) have remained intact (Wever, Neff, Schuknecht, Woellner).
2) After partial section of the VIIIth nerve, the hearing on the operated side may improve in the bass region (Aubry, Ombrédanne).
3) After partial section of the VIIIth nerve, a bass deafness may arise on the side that has not been operated upon (Aubry, Ombrédanne, Thiébaut and his associates).
4) After a streptomycin treatment of patients with a Ménière-like picture, owing to which the vestibular functions are entirely or partially lost, the hearing on one or both sides may improve in the bass region (Schuknecht).
5) An affection of the brain stem may lead to a bass deafness on one or both sides (Krassnig, Greiner and associates).
6) Partial section of the lateral lemniscus may cause a doublesided bass deafness (Walker, Saltzman).

How to bring all these findings onder one heading. More and more particulars have become known which point to it that the frequency localization, as found in the cochlea, is retained in the afferent acoustic system.

In this system, above the level of the cochlear nuclei, we find a division in the decussated and non-decussated nerve fibres.

From the experiments by Brogden, Girden, Mettler and Culler it has appeared that the non-decussated nerve fibres in the afferent central pathways have the same acoustic values as the decussated ones, for the low as well as the middle and high frequencies. Destruction of one cochlea of a cat causes a loss

of hearing of 3 to 4 dB. Ablation of a cerebral hemisphere on one side is followed by a loss of hearing of 3 to 5 dB. When the cerebral cortex of cats has been removed on one side, destruction of the homolateral or heterolateral cochlea causes an additional loss of hearing of about 15 dB.

So it may be thought very unlikely that a lesion of the afferent system, which is situated above the level of the brain stem, might cause a bass deafness of the type described by us. Moreover the six points summarized above could not be explained in case of such a lesion.

It is obvious that we should try and find the solution of this problem in the system of the efferent pathways, and more and more about these has become known to us during the last few years. For this it is necessary to give a short survey of the available data as to the course and the function of the efferent bundles of nerves in the central auditory pathways.

In the course of years the existence of important nerve pathways has been proved, which lead from the auditory cortex, via the various acoustic nuclei, to the cochlea. Thus there are connections between the cortex and the corpus geniculatum mediale, between the corpus geniculatum and the colliculus inferior and the nucleus of the lateral lemniscus, between the colliculus inferior and the superior olivary body on both sides, and between the superior olive and the dorsal cochlear nucleus on both sides. Finally there is a bundle of nerve fibres, originating from the ganglion cells in the neighbourhood of the superior olive, and lying close to the floor of the IVth ventricle, which joins the vestibular nerve from the other side; this is Rasmussen's olivocochlear bundle.

The fibres of this bundle leave the vestibular nerve near the saccular ganglion and form a vestibulo-cochlear anastomosis (Oort's bundle). They enter the spiral ganglion and terminate, according to some writers, on the inner hair cells of the organ of Corti; according to others this is not absolutely certain.

So something, though not much, is known about the function of the efferent system that has been outlined above.

Originally Rasmussen thought that the olivocochlear bundle consisted of vegetative nerve fibres which had a regulating influence on the blood vessels of the cochlea and the secretory epithelium of the stria vascularis. On the strength of this supposition he saw a close connection between affections of this bundle and the function of the auditory organ and perhaps also the vestibular organ (Ménière's syndrome).

Later on he altered his views and suspected a kind of feedback mechanism in the efferent system which was thought to have a regulating influence on the process of hearing. So a stimulation of the peripheral sense organ might be followed every time by an efferent stimulus back to the receptor.

This speculation is supported by the experiments made by Galambos, who showed that the action potentials in the auditory nerve of cats could be suppressed or could be made to disappear by means of electric stimulation of the floor of the IVth ventricle. These stimuli were applied to the crossing of the olivocochlear bundle. The phenomenon remained after removal of the ossicles and

muscles of the middle ear; it disappeared after section of the olivocochlear bundle peripheral of the place of stimulation.

GALAMBOS concludes from this that stimuli, led via the olivocochlear bundle to the periphery, can suppress the expected activity of the auditory nerve as a result of acoustic signals.

It is a pity that GALAMBOS used clicks as acoustic stimuli. This was the reason why he could not establish whether the suppression as described above, perhaps found particular expression in the regions of the low, the middle or the high frequencies.

Additional particulars are supplied by HERNÁNDEZ-PEÓN, SCHERRER and JOUVET. They describe the phenomenon of the habituation in the dorsal cochlear nucleus as a result of continually repeated acoustic stimuli. With a bipolar electrode the action potentials were measured in the dorsal cochlear nucleus while clicks were continually offered to the ear at intervals of two seconds. After some time the action potentials decreased or disappeared. This blocking effect was selective for the continually repeated stimulus; a new stimulus of a different character caused action potentials in the normal way. The blocking could be raised by a deep anaesthesia and by damaging the roof of the midbrain. Moreover the blocking for a certain stimulus could be called up by diverting the animal's attention from the stimulus and focusing its attention on something else.

The writers think this selective, descending, inhibitory influence must be attributed to the formatio reticularis in the brain stem. It may be considered just as likely that the efferent system which has been described above, must be held responsible for this.

With the help of the data which have been described so far, it is possible to draw up a scheme of the efferent system in the central auditory pathways. Figure 42 represents such a scheme; some of the afferent neurons have also been recorded.

We start from the supposition that the efferent system that has been described, has to exercise a regulating influence on the impulses entering via the afferent pathways. In view of the intricate structure of both the efferent and afferent systems, this regulating influence may take place at various levels. Thus, via the reflex arc cochlea – nucleus cochlearis ventralis – oliva complex – cochlea, which lies at a very low level, a reaction may follow immediately to impulses which enter the nucleus cochlearis ventralis via the cochlear nerve.

This low reflex arc is influenced by nuclei which are situated higher up and which are again connected with fibres of the afferent system.

We may ask ourselves the question what the regulating influence of the efferent system really is.

On the strength of the above-mentioned data we may obviously suppose that the task of the system is to suppress the impulses that have entered the afferent auditory pathways. This may be necessary if the impulse in question is unimportant or troublesome for the organism or if another impulse claims the full attention.

Diagram of the efferent auditory pathways

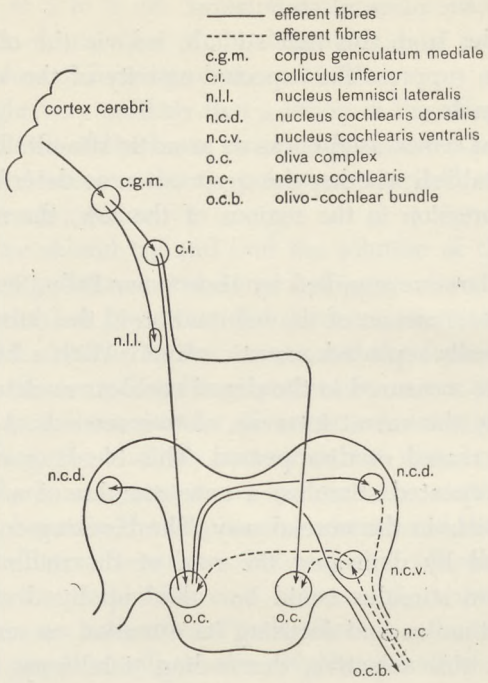


Figure 42

It must also be thought possible that a condition of hyperactivity in the efferent system itself may lead to suppression of afferent impulses, without being advantageous to the organism or even desired by it.

There is not a single experimental proof that the feedback mechanism that has been described above, would especially be active in the region of the low frequencies.

In the cochlear pathology no explanation could be found, however, for bass deafness in man of the type described, while it is not very likely either that a lesion of the afferent auditory pathways underlies it.

The strong and quick fluctuations of the hearing in the bass region (cf. our collection of patients), the mutual influence of the right and the left ear (Aubry, Ombrédanne, Thiébaut and his associates), the results of the streptomycin treatment of patients with a Ménière-like syndrome (Schuknecht), the occurrence of bass deafness in case of affections of the brain stem (Krassnig, Greiner and his associates) and the origin of a bass deafness on both sides in case of lesions of the lateral lemniscus (Walker, Saltzman) give us good reason to suppose that a bass deafness may arise under the influence of a central happening.

In view of the generally reversible character of the bass deafness (at least in the beginning), we get the impression that there is no question of irreparable destruction of the auditory pathways, but rather of a temporary elimination or suppression of the ability to perceive low tones.

We now suppose that the efferent system exercises a suppressive influence

on the afferent impulses and that this action finds particular expression in the region of the low frequencies.

What consequences may be expected from lesions at various levels in the auditory pathways?

A) Practically all our patients suffer more or less from tinnitus. It is imaginable that this tinnitus as an afferent impulse is able to invite a hyperactivity of the efferent system. This possibility should be all the more considered as this tinnitus is often of a low and buzzing character and thus the phenomenon of selective habituation of HERNÁNDEZ-PEÓN and associates might be evoked. It may depend on the place of origin of the tinnitus whether the bass deafness will be one or double sided.

B) If lesions are made in the cochlear nerve (SCHUKNECHT, WOELLNER), while retaining the vestibular nerve or at least Rasmussen's olivocochlear bundle, a bass deafness may arise. For this it is necessary that the efferent system is stimulated. This may take place in the meatus acusticus internus where, as a result of the interference, hemorrhage and oedema arise and later on perhaps scars. Perhaps LUNDBORG's case (acoustic tumour) may be compared with this. It may also be imagined that the cochlear nerve and the nucleus cochlearis ventralis will get into an irritated condition. In this case, a bass deafness may arise on both sides, owing to the afferent connections with the left and the right oliva complex. This has never been observed in the animal experiments described, but in these cases the cochlea was always destroyed on one side before the beginning of the experiments.

C) Section of the vestibular nerve with Rasmussen's olivocochlear bundle cannot result in a bass deafness on the side that has been operated upon (GUILD, WEVER, NEFF, AUBRY, OMBRÉDANNE). It is even like this that a possible bass deafness disappears on the homolateral side (AUBRY, OMBRÉDANNE). As a result of afferent connections between the nucleus cochlearis ventralis and the left and right oliva complexes a heterolateral bass deafness may arise in this case (AUBRY, OMBRÉDANNE, THIÉBAUT and his associates), as, in connection with the interference, there is always the possibility that the cochlear nerve and the nucleus cochlearis ventralis on the operated side will become irritated.

D) An encephalitis of the brain stem (KRASSNIG) may also cause a bass deafness owing to stimulation of the olivocochlear bundle and the nuclei of the efferent system. It depends on the place of the focus of the inflammation on which side the bass deafness will appear.

E) The same observations as under D hold good for affections of the IVth ventricle (GREINER and his associates). Here stimulation of the olivocochlear pathways may take place from the top. The chance that the bass deafness will be double-sided in these cases is to be considered great as the bundles from both sides cross very close to the floor of the IVth ventricle. This phenomenon was indeed found by GREINER.

F) A damage in the neighbourhood of the lateral lemniscus (WALKER, SALTZMAN) may lead to a stimulation of the efferent pathways which connect the

corpus geniculatum mediale and the colliculus inferior and via this nucleus with the left and the right oliva complex. A double-sided bass deafness may be the result of this.

Thus it seems possible to explain the origin of a bass deafness in a natural way from a condition of hyperactivity of the efferent system, from a surplus of feed-back.

We may wonder if all the phenomena found in our patients can be explained on the strength of this hypothesis.

In order to answer this question more or less satisfactorily it is necessary to discuss our own clinical material first, together with the methods of investigation applied. This will take place in the chapters VII and VIII. In chapter IX we shall revert to the question asked above.

Chapter VII

METHODS USED TO INVESTIGATE OUR PATIENTS

The anamnesis of each patient was taken down while special attention was being paid to the beginning of the complaints viz. whether these complaints had occurred suddenly or gradually. Enquiries were made about the hearing and about the degree and the nature of the tinnitus. If the patient had ever had disturbances in his equilibrium, he was asked to give a careful description of this. Diseases he had had before were noted down. The patient was asked if relatives had ever suffered from any hearing disturbances. Attention was also paid to the fact if the patient had ever been exposed to much noise or not and whether he had had an otitis media or a trauma capitis. Women were also asked if they had noticed that hearing complaints increased in connection with pregnancy. It stands to reason that all other particulars that became known during the anamnesis, were taken down, while finally attempts were also made to find a possible direct cause of the complaints.

A routine otorhinolaryngological examination was made of every patient. Siegle's otoscope was used to get a view of the ear drums and to check the mobility of them. Politzerization showed the degree of openness of the Eustachian tubes. A few times a diagnostic eardrum puncture was performed.

Threshold audiograms for pure tones were recorded in accordance with the method of octave audiometry. A Pedersen's audiometer was used with loudspeaker spheres. The loss of hearing was determined at the frequencies 125, 250, 500, 1000, 2000, 4000 and 8000 c.p.s. During octave audiometry a tone of a fixed frequency is offered every time, while with a gauged attenuator it is determined what intensity of tone can just be heard by the patient. This intensity indicates the loss of hearing, if compared with the intensity of tone needed by the normal ear so that it can just hear this particular tone. Many audiograms were made of each patient in the course of time.

Many times tone audiograms were recorded in accordance with the method of continuous audiometry. A Peekel's audiometer with a headphone was used for this. This audiometer, suitable for continuous audiometry, is provided with a continuously adjustable frequency scale. The frequency of the sound signal offered to the ear through the phone can consequently be varied over a frequency ranging from 125 to 8000 c.p.s. The sound level with respect to the normal threshold of hearing always remains the same when a certain reading of the attenuator is maintained. This sound level can be regulated by means of the attenuator with 5 dB at a time. This makes it possible to investigate the hearing so to speak in a horizontal direction (i.e. parallel to the normal threshold of

hearing) at various sound intensities. The patient indicates when he no longer hears the sound which varies continually in frequency or when this sound becomes audible again after having been inaudible. Thus various cross sections can be made of a dip-shaped loss of hearing and the form, size and location can be recorded perfectly by scanning closely. The method of continuous audiometry is pre-eminently suited to discover something about the real steepness of the curve of an audiogram. In Holland spade-work was done by VAN DISHOECK to develop this method and to make it fit for use in practice.

If possible a speech audiogram was made. Wordlists of monosyllables are used in speech audiometry; good care is taken that the various sounds occur in them in the same proportions as in normal speech (the so-called phonetically balanced wordlists). The words are offered to the ear which is to be examined by means of a loudspeaker mounted in a sphere. The words have been recorded on a magnetophonic tape at a constant intensity of speech and are played back via a gauged attenuator. At various readings of the attenuator it is determined what percentage of the words is understood correctly. At reading 0 of the attenuator 50 % of the words are understood correctly by the normal ear. The data thus obtained are rendered in a diagram. On the horizontal axis we can read at what intensity of speech 50 % of the words are correctly understood. This indicates the loss of hearing. On the vertical axis we can read what percentage is maximally understood at optimal amplification.

Several times a noise audiogram was made, in accordance with LANGENBECK's method. According to this author this method makes it possible to distinguish between two kinds of deafness: the inner ear or hair cell type and the ganglion or nerve type. In the first case a white noise is supposed to have a masking effect which is as great as that in a normal ear, also in those regions in which an increased threshold exists. In the second case a white noise is supposed to have an abnormally great masking effect in those regions in which there is an increased threshold. In the latter case the tones must be much louder than the noise to be audible.

Various investigators have tried to find an explanation for this phenomenon. Some writers (LÜSCHER and ZWISLOCKI, 1949) thought that the masking came about because of the adaptation of the hair cells. The noise causes the hair cells to adapt, and the result is a reduced irritability. Therefore, when a tone stimulus is added, the hair cells can only then be stimulated when it is louder than the noise, in other words the tone stimulus must at least be equal to the noise stimulus. Only then does the tone stimulus in the inner ear cross the threshold of the hair cells which are adapted to the noise, no matter whether we have an inner ear with normal hair cells or an abnormal inner ear with abnormal or a reduced number of hair cells. But what about the disturbances in the nerve? Let us presume we have a real disturbance of the nerve. Then we can imagine that in spite of normal irritability of the hair cells, the inner ear would have to be stimulated beyond the (normal) threshold in order to make it at all possible to lead a stimulus along the defective auditory nerve to central. The inner ear must

for instance be stimulated 20 dB above the (normal) threshold before the nerve becomes active at all. When the tone stimulus and the noise stimulus are equally strong, the tone stimulus, according to the theory, will overstep the threshold of the hair cells that are adapted to the noise. So in case of an impaired auditory nerve the tone stimulus must be considerably greater than the noise stimulus in order to be able to supply 20 dB extra, which are necessary to make the nerve conduct the stimulus. From this theory it follows that in case of a nervous disorder the hearing of tones in noise must be worse than in a normal ear or an ear with a hair cell disturbance.

Other writers (STEVENS and DAVIS, 1948) see the cause of the masking phenomena in the connection of the hair cells with the nerve fibres or rather the dendrites of the spiral ganglion. In consequence of the refractory period of the cells of the spiral ganglion, the stimuli are not conducted any further when the ganglion cell has received a stimulus shortly before. Every ganglion cell only then becomes irritable again, when its refractory period (about 1 msec.) is over. As a result of the numerous connections, conditions arise which may be compared to those in a telephone system. According as the traffic in such a system becomes more intensive, it happens more often that in spite of numerous transverse connections a certain call cannot be put through. The connections which cannot be established or the calls which do not come through correspond with the masked tone stimuli. Only a sufficiently strong tone stimulus, so a stimulus which activates a sufficiently large number of hair cells, has a statistical chance to penetrate into the auditory nerve together with an existing noise stimulus. The proportions must become more unfavourable when nerve fibres drop out owing to pathological processes. In case of a disturbance in the auditory nerve, the tone stimuli must consequently be stronger than the noise stimuli in order that centrally the perception of tone may come about together with the perception of noise.

A noise audiogram is made as follows. First the threshold audiogram for pure tones is determined. Then this is repeated but now at the same time a white noise is offered to the ear to be examined, the masking influence of which on the normal auditory organ is known. In the ultimate diagram three curves are to be seen: the threshold audiogram for pure tones, the masking audiogram of the white noise for the normal auditory organ and the threshold audiogram for pure tones in the presence of the white noise. Now we can see at a glance whether the noise had a masking effect which was normal or abnormally great.

For the recording of the threshold audiogram for pure tones in accordance with the methods of octave audiometry and of continuous audiometry, as well as for the recording of speech and noise audiograms, the normal ear was masked when the abnormal ear was examined.

As the bone conduction curve in our audiogram always coincided with the air conduction curve of the corresponding ear (bone conduction of the abnormal ear was always recorded under simultaneous masking of the normal ear), the latter is not rendered in the audiograms shown.

By diplacusis we understand the phenomenon that the same tone, offered

successively to the left and the right ear of the same person, seems to have a pitch in one ear different from that in the other (diplacusis binauralis dysharmonica). This phenomenon is practically always present to a slight extent in normal cases. In pathological cases it may be very pronounced, even to such an extent that the patients are greatly troubled by it when listening to music. Most writers (SHAMBAUGH among others) try to find the cause of this phenomenon in a disturbance of the normal hydrodynamic relations in the cochlea, owing to which the maxima of the stimulation patterns of the basilar membrane would be in places different from the normal ones. This might be the case for instance when the basilar membrane would be made heavier by inflammatory exudations and if thus a change in mass and stiffness of this membrane would come about. A tone, offered to the ear, is then no longer perceived by its own hair cells but by hair cells which are properly speaking tuned to another frequency, which frequency is then taken for this tone. This explanation fits in completely with the place theory in its most stringent form. During Fowler's balance test the patients were mostly also asked how they rated the tones that were in turn offered to both ears, in the abnormal ear with respect to the normal ear, i.e. higher, lower, impure, etc.

The term recruitment in audiology is given to the phenomenon of a pathological increase of the impression of loudness when the intensity of the tone offered to the ear increases. If a tone is offered with an intensity of for instance 20 dB above the threshold, it is heard louder by a patient with recruitment than by a patient whose deafness is not characterized by recruitment or by somebody with normal hearing. Often the phenomenon is so pronounced that finallly the impression of loudness at higher intensities of tone is completely equal to that of a person with normal hearing. This has been proved in patients who had a normal hearing on one side and a hearing loss with recruitment on the other side. There are many methods to establish the presence of the recruitment phenomenon. In cases of unilateral deafness we can still best avail ourselves of the technique, originally introduced by FOWLER, the so-called alternate binaural loudness balance test. A tone, of the frequency for which we want to establish whether there is recruitment or not, is offered to the normal and the abnormal ear alternately. The intensity of this tone is gradually increased. In the beginning the tone is only heard in the normal ear; it remains like this until the threshold of the abnormal ear is overstepped. From this moment onwards the patient has to indicate every time when the tone causes an equal impression of loudness in both ears; this is repeated at continually increasing intensities and the intensities at which this occurs are noted down and rendered in a diagram. In this diagram we can see the difference in hearing thresholds between the two ears for a certain frequency and the way in which the abnormal ear makes up for its arrears. Most writers consider the recruitment phenomenon to be an important expedient in diagnostics and they even see the phenomenon as pathognomonic for affections of the organ of Corti, whereas they say that it does not occur with lesions of the VIIIth nerve and the higher acoustic pathways. There are other investigators

(GREINER among others) who point out again and again that they have found the recruitment phenomenon in cases of hearing disturbances which were a result of affections of the central nervous system. If possible our patients were examined according to FOWLER's method.

Some ten patients were examined by our colleague SEDEE. This investigator showed that the time relation of a vibration at the left and the right stapes footplate is the most important parameter for directional hearing. As the small differences in time between moments of arrival of the vibrations on the left and the right eardrum are preserved during neural transport in the first neuron, the moments of arrival of the action potentials in the acoustic nuclei have the same differences in time. The hypothesis that there is a region in the medulla (the corpus trapezoideum), at the first meeting of the auditory pathways from the left and the right ear, where the correlation of the stimuli from both ears takes place, could be confirmed by the experiments with patients suffering from syringobulbia, whose directional hearing had been seriously disturbed or had got lost, while the threshold audiograms for pure tones did not show any abnormalities. In the brain stem the foundations for stereo-acoustic perceptions are laid. By means of the method developed by SEDEE to determine the accuracy of directional hearing, we have tried to get data for a further localization of a possible lesion of the central acoustic system.

The function of the organs of equilibrium were examined by means of the caloric test, the turning chair and mostly also by means of the marche en étoile and the Unterberger's and Romberg's tests.

The caloric test is carried out as follows. The patient lies on his back on a level bench, the top of which is elevated 30°. The chin is consequently bent a little towards the chest; the horizontal semicircular canals are now vertical. The room in which the examination takes place is in twilight and the patient has moreover Frenzel's glasses (+ 20 diopters) which can be lighted on the inside. Next in the course of 20 seconds 100 cc water of 44° Celsius is syringed into the external auditory canal of the right ear and the resulting nystagmus is timed with a stopwatch. At the same time attention is paid to the quality of the nystagmus. Then this is repeated on the other side. Finally the right and the left ear successively are syringed under the same circumstances with water of 30° Celsius, while attention is also paid to the duration and quality of the nystagmus. The four values thus obtained are rendered in a diagram and thus it is shown immediately whether both labyrinths are equally excitable (equal values on both sides for water of 44° and equal values for water of 30°), whether there is a predominance of one labyrinth (higher values on one side for both temperatures than on the other side), or whether we have the phenomenon that the nystagmus preferably takes a certain direction (nystagmus-preponderance). The latter phenomenon for instance presents itself if the longest duration of the nystagmus is found when water of 44° is syringed on the right side, while the nystagmus on the left side lasts longest with water of 30°. Then we have a preponderance to the right.

During the turning chair examination the patient is put in an electric turning chair, also in twilight and wearing lighted Frenzel's glasses. The head is bent forward 30° so that the horizontal semicircular canals are really horizontal. Then the turning chair is turned round with a subliminal acceleration, until the velocity required (angular velocity) is reached. In this way no perrotatory reactions will occur. After having rotated for some time at the velocity reached, the chair is suddenly stopped. Up to that moment the patient has not had any gyratory sensations and any form of nystagmus has been absent. At the moment of stopping he will get a gyratory sensation, however, and that opposite to the real direction of rotation. This gyratory sensation will gradually decrease and finally the patient will think he has come to a standstill. Before the examination he has been requested to communicate this moment to us. In this way the duration of the after sensations for both directions of rotation at a number of different velocities is determined.

Besides a gyratory sensation when the patient comes to a standstill, a nystagmus arises with the fast phase to the original direction of rotation. The duration of the postrotatory nystagmus is measured in the same way as this is done for the sensations. The best results are obtained if the duration of the after sensations and the duration of the post-rotatory nystagmus are measured by determining separate progressions.

When the uniform rotatory movement is suddenly stopped, the cupula turns off as a result of the inertness of the endolymph. The cupula, as a result of its elastic qualities, straightens itself again until equilibrium is reached. During this time postrotatory phenomena arise. From the calculations (GROEN) it appears that the length of time is proportional to the logarithm of the impulse causing the postrotatory reaction. The impulse value of this is determined by the magnitude of the uniform velocity of rotation before stopping. The magnitude of the impulse is consequently expressed in that of the angular velocity. The values obtained with the turning chair examination are therefore plotted in a logarithmic diagram. The curves connecting the points determined, are generally rectilinear.

During our examination we confined ourselves to the measurement of the duration of the postrotatory nystagmus.

By marche en étoile we understand walking forwards and backwards repeatedly with closed eyes in a room in twilight. Unterberger's test is a simpler form of this viz. the patient lifts his right and his left leg consecutively without walking and with his eyes closed. In both cases we want to establish whether, under the circumstances mentioned, there is a deviation to the left or the right, in other words whether the patient turns on his axis during the examination. Romberg's test is performed with eyes closed and the feet close together, the patient remaining like that for some time. It is checked whether the patient shows any tendency to fall to some side or other.

In principle we may expect that in case of a reduced function of for instance the left labyrinth, the patient will revolve to the left round his axis during the marche en étoile and the Unterberger's test. During Romberg's test the patient

will have a tendency to fall to the left. For the rest no absolute value may be attached at all to the results of the tests described, as these results are highly dependent on the varying influences of compensation mechanisms and tonus relations during the course of the disease. In cases of recent affections of the labyrinth the results are most reliable.

If possible our patients were subjected to a general neurological examination. Unfortunately it was not possible to make an electro-encephalogram in all the cases.

In most cases roentgenograms were made of skull and mastoids (transverse and fronto-occipital photos of the skull and photos according to STENVERS, STEENHUIS, SCHÜLLER and MAYER).

If necessary the patient was examined internally. Some patients were examined ophthalmologically.

Chapter VIII

CASE HISTORIES

Forty-five patients with bass deafness were examined by us, 25 men and 20 women. During the first examination the tone audiogram was 18 times of type I, 8 times of type II, 15 times of type III and 4 times of type IV (cf. the introduction). We may observe here that the distribution over the various types has not been of essential importance. During an observation period ranging from some months to some years, a transition from type I to type II was seen 7 times. Patients with a type III audiogram mostly had a more or less pronounced presbycusis, while patients with a type IV audiogram had always worked in noise. We shall first give a survey of the findings in our collective material, and then proceed to discuss each type separately and to describe some representative cases of these types.

Most patients came to us when they had had hearing complaints for some time already. The anamnesis showed at what age the complaints had started. In the table given below a survey of this is given.

Table I

age when complaints started	number of patients total 45
0–10	2
11–20	2
21–30	8
31–40	11
41–50	10
51–60	8
61–70	3
71–80	1

In 34 cases the hearing abnormalities were unilateral (13 times left, 21 times right); a bass deafness of one of the types described was always found in these cases. In 11 cases the hearing of the other ear was impaired as well; while there was a bass deafness in one ear, the audiogram in some cases showed a bass deafness for the other ear as well, but mostly the nature of the hearing loss was flat.

In the anamnesis 14 patients mentioned possible starting points for the question of the origin or the occasion of complaints. These were: once a cold, once influenza, once otitis media, once chronic rhinosinusitis, once herpes zoster, once anemia, once explosion of fireworks, once trauma capitis, once a streptomycintreatment in case of infection of the lungs, 4 times familial deafness while once the bass deafness was said to have been congenital.

An internal examination revealed: twice an anemia, twice a hypertension, one of these cases being attended with a gallbladder disease, once slight gallbladder complaints and once a bronchial asthma with emphysema and fibrosis of the lungs and attended with a bad functioning of the kidneys.

An ophthalmic examination revealed 3 times a third stage arteriolosclerosis in the fundus, once attended with a glaucoma.

A routine otorhinolaryngological examination revealed that abnormalities of the ear drums did not occur at all. In all the cases the Eustachian tube was patent. A few times an ear drum was punctured from diagnostic considerations; this was always negative.

Roentgenological examination never showed any abnormalities in the temporal bone; photos according to Schüller, Mayer, Stenvers and Steenhuis were made in many cases.

A tone offered alternately to the nonaffected and the affected ear, was in most cases appreciated by the second ear in a way different from the first. In the affected ear the tone sounded higher or lower than in the non-affected ear, while the perception of it was often distorted or impure. This phenomenon – diplacusis – was practically always found.

On investigation it appeared that there was also a recruitment phenomenon in all cases. Sometimes, it is true, the phenomenon was not pronounced and could often hardly be proved, but in as many cases it was pronounced and could be fully proved.

In 15 cases noise audiograms were made, which were always more or less of the ganglion type.

Thirty-five times there was a general neurological examination. In 15 cases neurological abnormalities were found; in the other 20 cases no neurological abnormalities could be found even when the examinations were repeated later on. The neurologist diagnosed tentatively one neuritis of the Vth nerve and the VIIIth nerve, once a facial hemiatrophy and nuclear atrophy, once an affection of the vegetative nervous system, once arachnoidal adhesions after trauma capitis, once a brain stem syndrome, once a general arteriosclerosis; three times there were abnormalities in the pyramidal tract but these abnormalities could not be co-ordinated with the rest of the findings while 6 times solitary neurological symptoms were found, the importance of which was to be considered doubtful.

An electro-encephalogram was made in 23 cases. In 16 cases there were abnormalities, mostly isolated or diffuse paroxysmal epileptiform activity.

In all the cases a vestibular examination was made (calorigram mostly supplemented by a cupulogram). Twelve patients did not complain of vertigo and

a vestibular examination did not reveal any abnormalities. Five other patients did not complain of any disturbances in the equilibrium either but in these cases the examination did show abnormalities. Eight patients told that they were troubled by disturbances of the equilibrium, but a vestibular examination did not show any abnormalities of the labyrinthine function. Finally a group of 20 patients complained of vertigo while the examination did show abnormalities of the labyrinthine function.

In 10 cases our colleague SEDEE made experiments to determine the accuracy of directional hearing. In 9 patients no abnormalities were found in this respect, at least if the hearing loss in the bass region was not more than about 60 dB. If the hearing loss in this region was more than 60 dB, the low tones that were offered to the affected ear, were also perceived in the non-affected ear (over-hearing!) owing to which some of the accuracy of the directional hearing was lost. The directional hearing for high tones was always unimpaired in these 9 patients. Only the directional hearing of one patient (case 3) was impaired for all frequencies; while only a hearing loss of 20 dB was found in the threshold audiogram for pure tones for both ears in the frequency region from 2000 up to and including 8000 c.p.s., the patient could only determine with great difficulty from what direction the tones of 2000 and 4000 c.p.s. came. It was assumed that in this case at medullar level there was a disturbance in the possibilities of interaction of both auditory organs.

TYPE I

In our material there are 18 patients with a bass deafness of type I. Twelve of these had a unilateral deafness (5 times on the left side, 7 times on the right side), while in 6 cases the hearing of the other ear was also impaired. As was stated already in the beginning of this chapter, we saw a transition from type I to type II in 7 cases during an observation period ranging from some months to some years.

The average age at which the patients with the unilateral hearing impairment were examined by us for the first time was 35 (varying from 16 to 63). The average age at which the complaints had started was 34 (varying from 16 to 63). The complaints had an average duration of one year (varying from 2 weeks to 6 years).

The average age at which patients with the bilateral hearing impairment were examined by us for the first time, was 39 (varying from 19 to 57). The average age at which the complaints had started was 28 (varying from 0 to 57). The complaints had an average duration of 11 years (varying from 3 weeks to 34 years).

Thirteen patients of this group were subjected to a general neurological examination. In 4 cases the examination revealed abnormalities. The neurologist established the following tentative diagnoses, once a neuritis of the Vth nerve and the VIIIth nerve, once a facial hemiatrophy with nuclear atrophy, once an

affection of the vegetative nervous system, while once abnormalities were only found in the pyramidal tract, which did not give rise to any diagnosis.

In 10 cases an electro-encephalogram was made; abnormalities were found in 6 cases. A vestibular examination was made of the 12 patients with a unilateral deafness. Four of these, who had no complaints of disturbances of equilibrium, did now show any abnormalities. One patient, who did complain of vertigo, did not show any abnormalities either. In 2 patients who had no complaints as to their equilibrium, a predominance of the labyrinth on the non-affected side was found twice during the turning chair examination (once this result was not confirmed by the caloric test). The caloric tests revealed that of the 5 patients who did complain of vertigo, 3 showed a predominance of the labyrinth on the non-affected side (once not confirmed by the turning chair examination), one had a preponderance to the non-affected side (not confirmed by the turning chair examination) and one had a preponderance to the affected side.

The same examination was performed with the 6 patients who also had an abnormal hearing on the other side. Three of these, who had no equilibrium complaints, did not show any abnormalities. The caloric tests revealed that one patient, who did not complain of vertigo, had a predominance of the labyrinth on the side of the bass deafness (not confirmed by the turning chair examination). In 2 patients who did have equilibrium complaints, once a predominance of the labyrinth on the side of the bass deafness was found (perhaps we may also call it a preponderance to this side) and once a preponderance to the side of the bass deafness.

Case 1

H.B. man, born 1914, elementary school teacher.

The patient was examined by us for the first time on 23-4-1957. At that moment he had been troubled by tinnitus in the right ear for 2 years. His hearing on the right side had become less good. The patient complained of a feeling of fullness in this ear. There were no complaints of vertigo. Deafness did not occur in the patient's family. The patient had never been ill. The hearing in the right ear was according to the patient, of a varying nature i.e. sometimes a little better, then again worse.

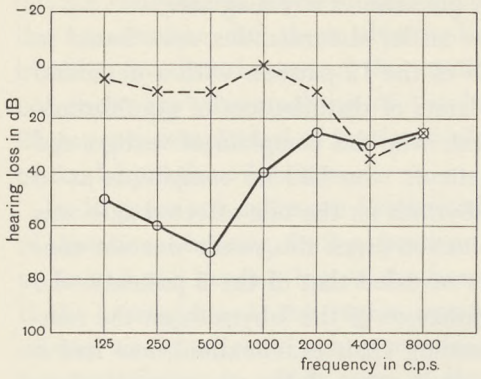
The routine otorhinolaryngological examination did not reveal any abnormalities. No abnormalities were found in the roentgenograms of the temporal bones (made according to Schüller, Mayer and Stenvers). A general neurological examination did not show any abnormalities either.

The electro-encephalogram showed some diffuse beta-activity. There was some diffuse theta activity, predominantly in the temporal region, a few times on the left more than on the right. Response to photic stimuli. Conclusion: no clear abnormalities.

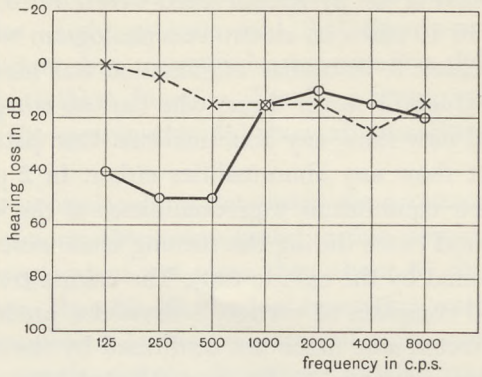
The vestibular examination showed a predominance of the left labyrinth both in the calorigram and the cupulogram. During the marche en étoile and the tests of Romberg and Unterberger no abnormalities were found, however.

The threshold audiogram for pure tones of 23-4-1957 showed a pronounced bass deafness in the right ear. A follow-up examination on 29-7-1957 revealed that the hearing had inproved a little. On the same day a continuous threshold audiogram was made too. The Fowler's test (binaural balance) showed the presence of recruitment in the right ear at 500 c.p.s. In the speech audiogram there was a hearing loss of only 20 to 25 dB, while speech perception was 100 % when sufficiently amplified (figure 43).

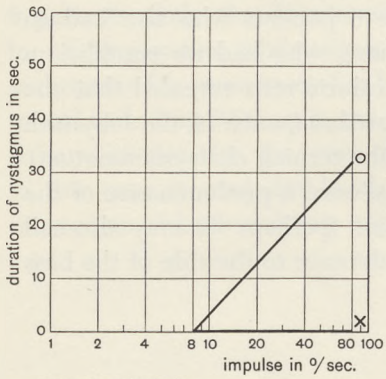
Date 23 - 4 - '57 Octave audiogram



Date 29 - 7 - '57 Octave audiogram



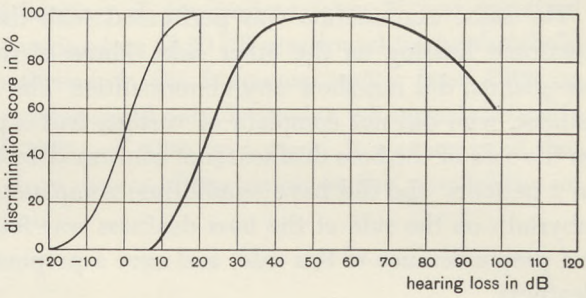
Date 29 - 7 - '57 Cupulogram



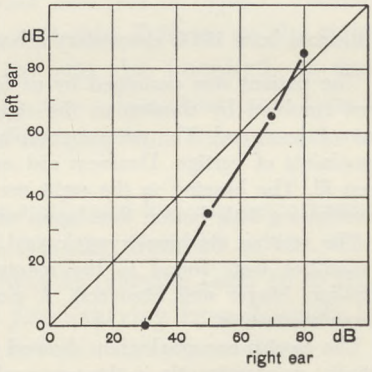
o turning clockwise

x turning anti-clockwise

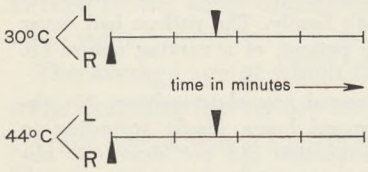
Date 29 - 7 - '57 Speech audiogram



Date 29 - 7 - '57 Loudness balance



Date 29 - 7 - '57 Calorigram



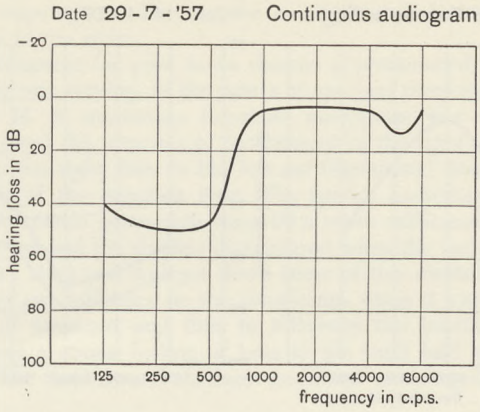
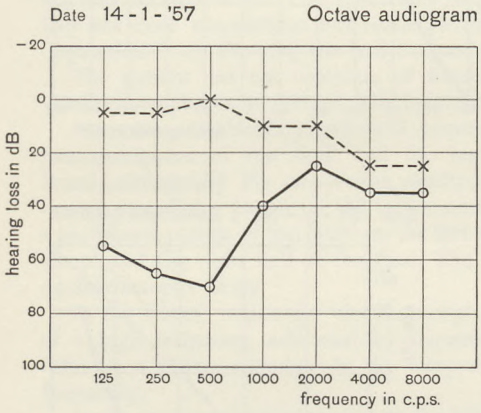


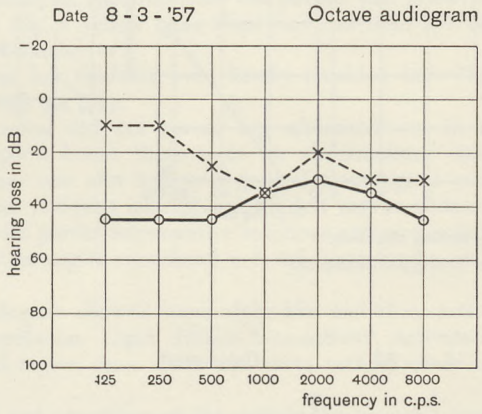
Figure 43

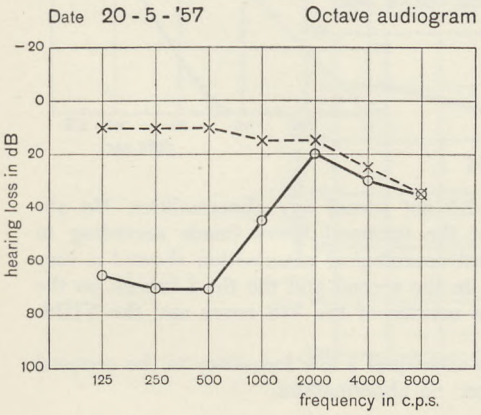
Case 2

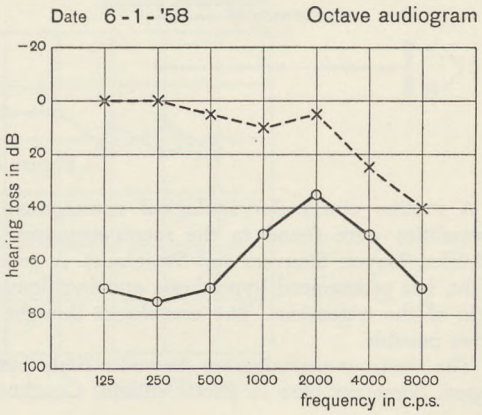
A. v. d. E., woman, born 1915, chemist's assistant.

The patient was first seen by us on 17-12-1956. She had had complaints of tinnitus in the right ear for 4 weeks. At that moment her hearing was not so good either. Patient also complained of slight dizziness; she was giddy but there was no objective vertigo. All this had cropped up after a cold.









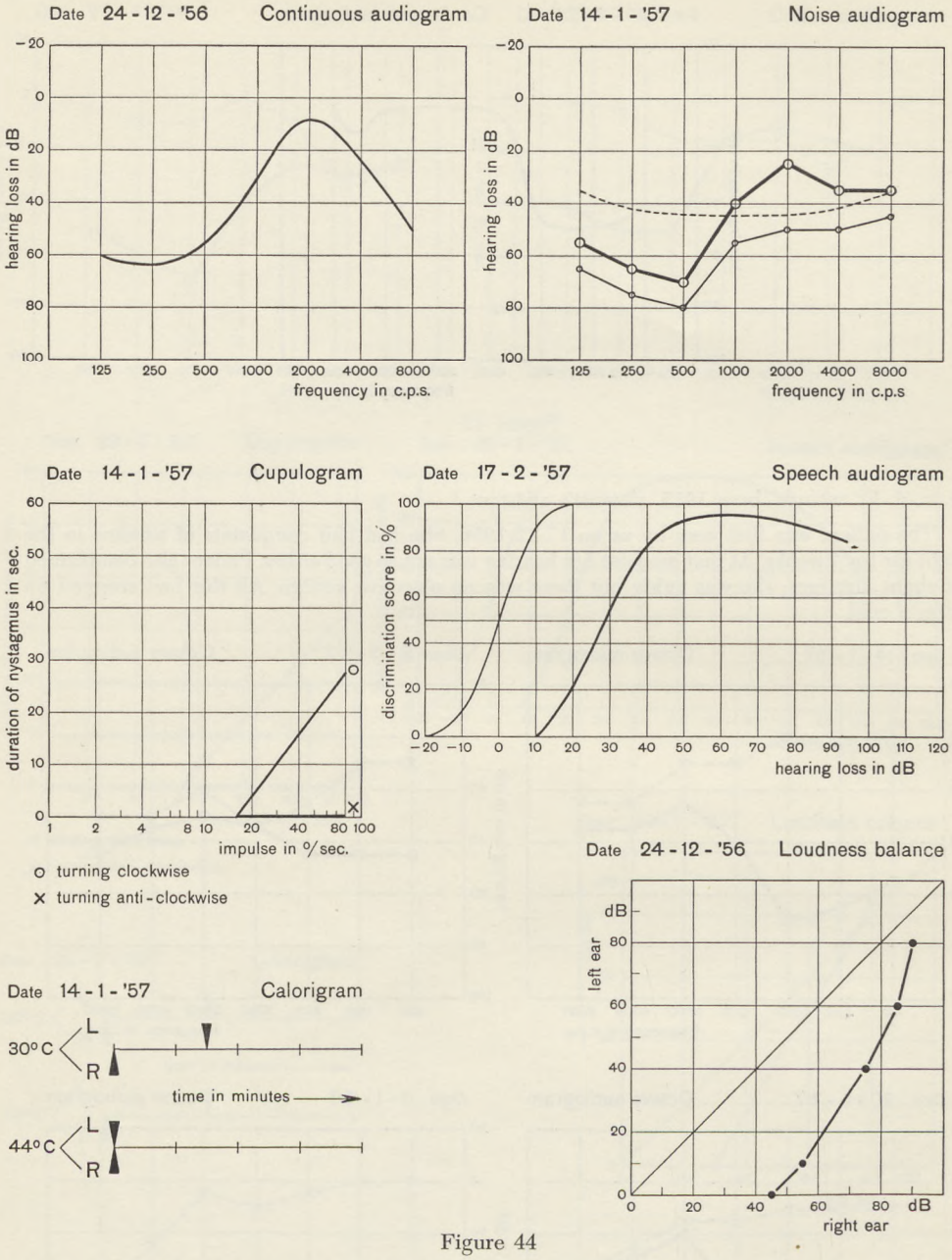


Figure 44

A routine otorhinolaryngological examination did not reveal any abnormalities. No abnormalities were found in the roentgenograms of the temporal bones (made according to Schüller, Mayer, Stenvers and Steenhuis). A general neurological examination showed a very slight, but pronounced hypesthesia and hypalgesia in the second and the third branch on the right of the trigeminus. The neurologist thought a neuritis of the Vth nerve and the VIIIth nerve possible.

The electro-encephalogram showed a slight beta-activity of a low frequency in the occipital region. Some response to photic stimuli. Conclusion: no abnormalities.

The vestibular examination revealed a predominance of the left labyrinth both in the calorigram and the cupulogram. The marche en étoile and Unterberger's test proved that there was a deviation to the right.

The threshold audiograms for pure tones showed a pronounced bass deafness in the right ear; the loss of hearing was varying. In the course of one year there was a clear tendency towards a transition to type II. A continuous threshold audiogram was made once. Fowler's test (binaural balance) showed the presence of recruitment in the right ear at 250 c.p.s. The tones had a higher pitch in the right than in the left ear (diplacusis), but did sound pure. Langenbeck's audiogram was of the ganglion type. The speech audiogram revealed a hearing loss of about 25 dB, while speech perception was 100 % when sufficiently amplified (figure 44).

The hearing improved and the tinnitus disappeared when the patient had prepared Largactil tablets at the chemist's shop and had got down some of this medicine. A Largactil treatment, however, did not have any influence on the complaints. Once it was tried to block the stellate ganglion by means of novocain and thus to influence the hearing. Though a pronounced Horner's syndrome and a strong feeling of heat in the right half of the face and right neck region were evoked, the tinnitus and deafness remained unchanged.

Case 3

T. de G.-v. H., woman, born 1924, housewife.

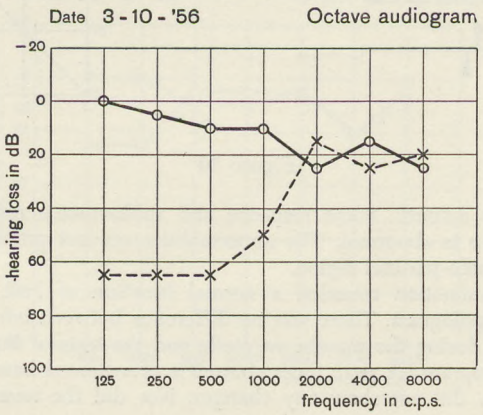
The patient was for the first time examined by us on 9-1-1956. Eight years before she had noticed that the left corner of her mouth began to droop; this was progressive, while moreover the paralysis spread over the whole of the left part of the face. Since 2½ years she had seen double images when looking to the left and upward. For some months the hearing of her left ear had become worse. The syndromes of paralysis occurred after the patient had fallen; she had not been unconscious but remembered that a yellow clear fluid had run from her nose (liquorrhea?). Swallowing too had become difficult.

The patient did not complain of tinnitus, her deafness even hardly troubled her. There were no complaints of vertigo. The hearing did not vary.

A routine otorhinolaryngological examination did not reveal any abnormalities. In the roentgenograms of the skull and the temporal bones there were no abnormalities either. Roentgenologically the swallowing mechanism was also intact. A general neurological examination revealed a paresis of the left musculus obliquus inferior and the left nervus abducens, a peripheral paresis of the facial on the left with partial degenerative reactions and a remarkable atrophy of the right half of the face. The neurologist considered a facial hemiatrophy and a nuclear atrophy likely.

In the frontal region the electro-encephalogram showed some alpha-like and theta activity of a high frequency activated by hyperventilation. Light diffuse beta-activity, activated a little by a photic stimulus. In the temporal region some delta and theta activity of a high frequency.

In the temporo-parietal region on the left and sometimes on the right in the temporal region



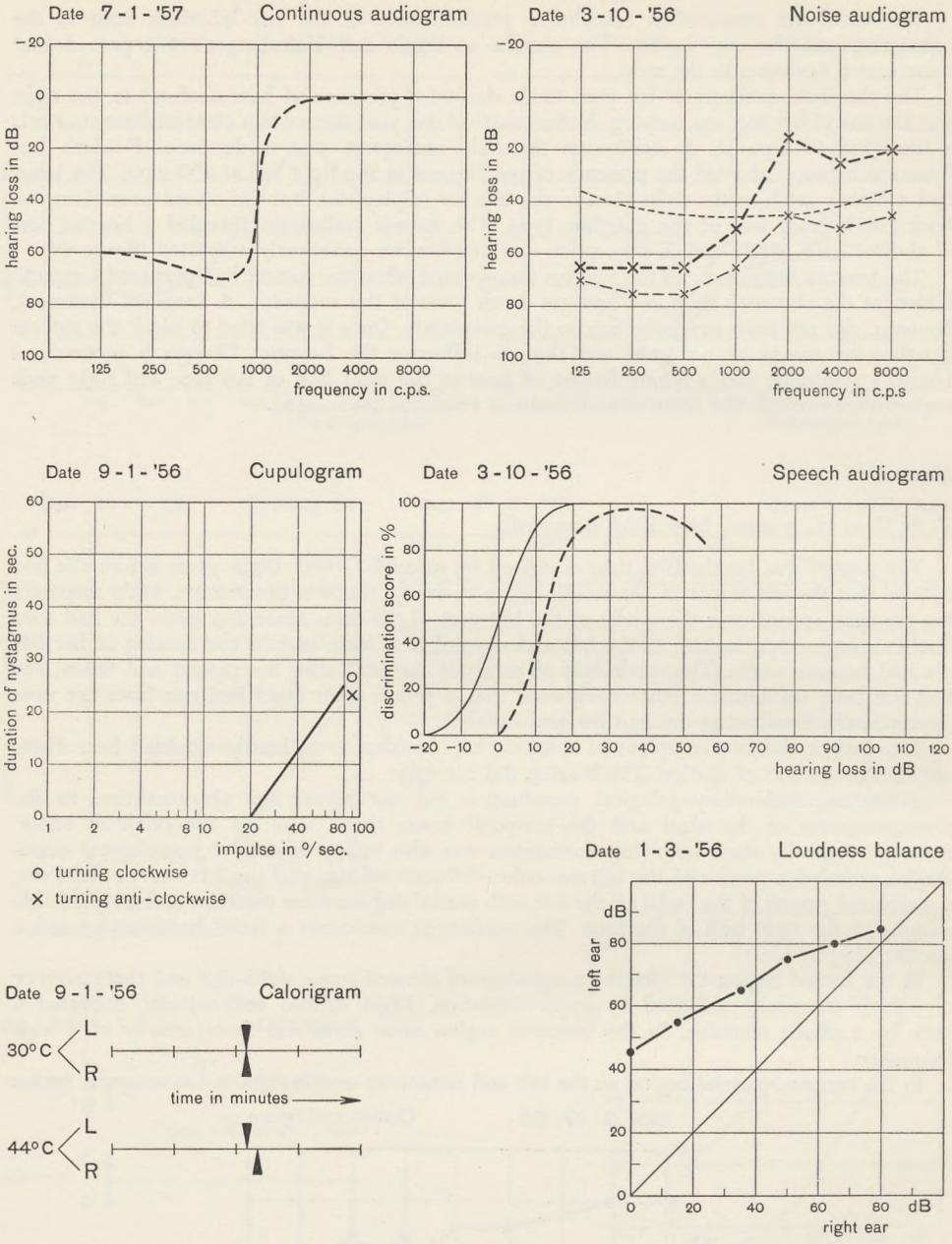


Figure 45

there was some spike activity. Some response and subharmonic response to photic stimuli. Conclusion: the picture is abnormal. The abnormalities are not pronounced and predominate on the left in the temporo-parietal region.

The vestibular examination revealed a normal function of both labyrinths both in the calorigram and the cupulogram. There was no difference between left and right. No abnormalities were discovered during the marche en étoile and the tests of Romberg and Unterberger.

The threshold audiogram for pure tones showed a pronounced bass deafness in the left ear. Follow-up audiograms did not show any changes. Nor did the hearing vary subjectively. A

continuous threshold audiogram was also made. Fowler's test (binaural balance) proved the presence of recruitment in the left ear at 500 c.p.s. Langenbeck's noise audiogram was of the ganglion type. The tones were of a higher pitch in the left ear than in the right ear (diplacusis) but they did sound pure. In the speech audiogram there was a hearing loss in the left ear of only 15 dB, while speech perception was 100 % if sufficiently amplified. During the tests to establish the accuracy of directional hearing (our colleague Sedee) gross abnormalities were found, also for pure tones of 4000 c.p.s. The patient could only determine with great difficulty from what direction a sound came (figure 45).

Case 4

A. M.-R., woman, born 1917, housewife.

The patient was seen by us for the first time on 9-11-1955. For 6 years she had been troubled by noises in the head; since a year tinnitus of a low nature in the right ear had been added to this. She had a feeling that her right ear was blocked. Loud sounds were experienced as disagreeable. There were no complaints of vertigo. Patient had been treated with liver injection some time before because of an anemia. The complaints had started after this, according to her.

The routine otorhinolaryngological examination did not reveal any abnormalities. An ear drum puncture was negative.

The roentgenograms of the temporal bones (according to Stenvers and Steenhuis) did not show any abnormalities.

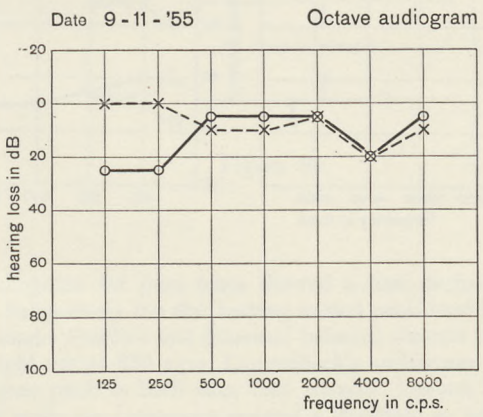
A general neurological examination did not show any abnormalities either.

The electro-encephalogram showed an alpha-rhythm of a low frequency, activated a little by hyperventilation. Diffuse beta-activity predominantly in the temporo-parieto-occipital region. Some theta activity in the temporo-parieto-occipital region, sometimes a little more on the left than on the right. Some response to photic stimuli. In the parieto-occipital region some slight spike and spike-and-wave like activity. Conclusion: irregular picture. The response to flash stimuli is slightly abnormal.

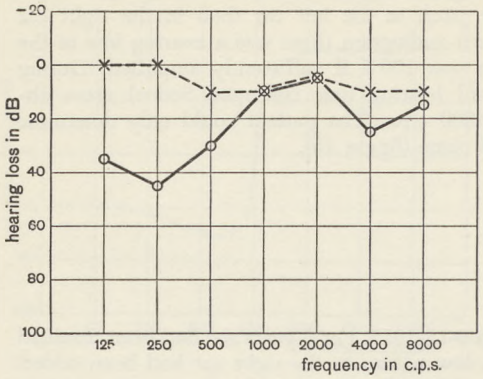
A general internal examination revealed a slight systolic souffle at the apex of the heart and also a low hemoglobin percentage of the blood (68 %).

The vestibular examination showed a normal function of both labyrinths both in the calorigram and the cupulogram; there was no difference between the left and right. No deviation during the marche en étoile.

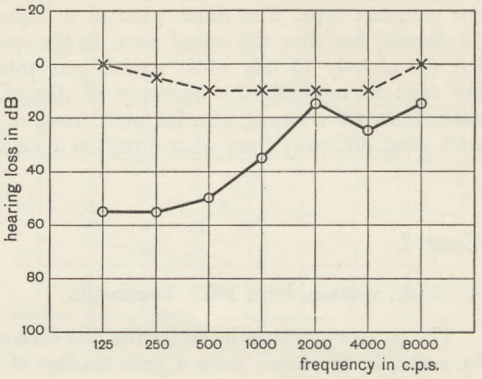
Nearly two years later the patient did complain of light fits of vertigo (10-9-1957). By means of the caloric test a perfect nystagmus could be caused on both sides. Romberg's test caused the patient to fall backwards and to the right. During the marche en étoile and Unterberger's test there was a deviation to the left.



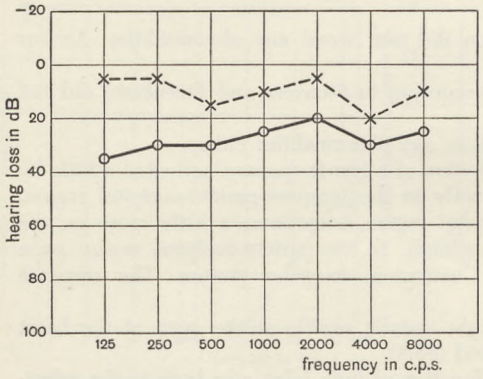
Date 12 - 1 - '56 Octave audiogram



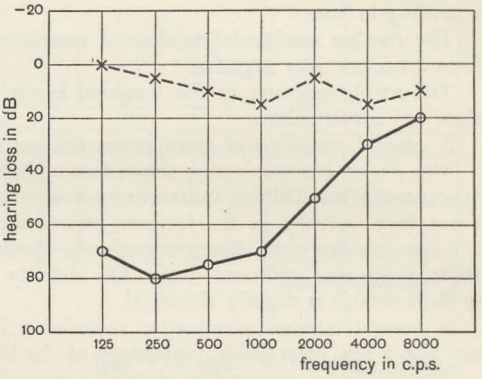
Date 18 - 1 - '56 Octave audiogram



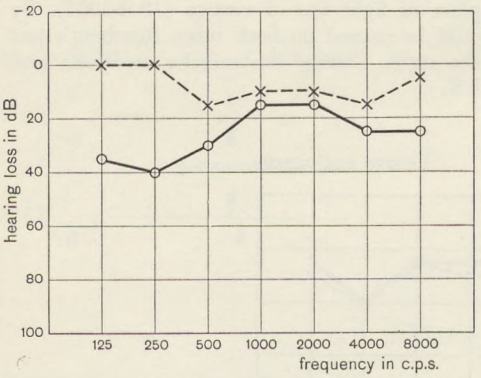
Date 7 - 2 - '56 Octave audiogram



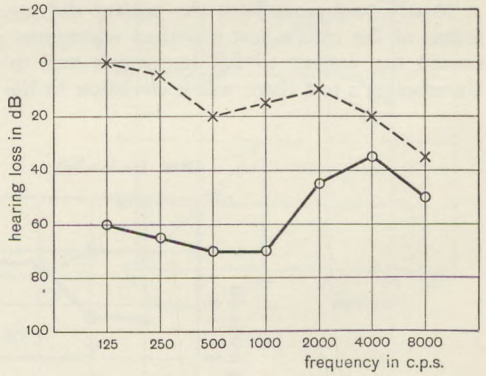
Date 5 - 7 - '56 Octave audiogram



Date 16 - 8 - '56 Octave audiogram



Date 10 - 9 - '57 Octave audiogram



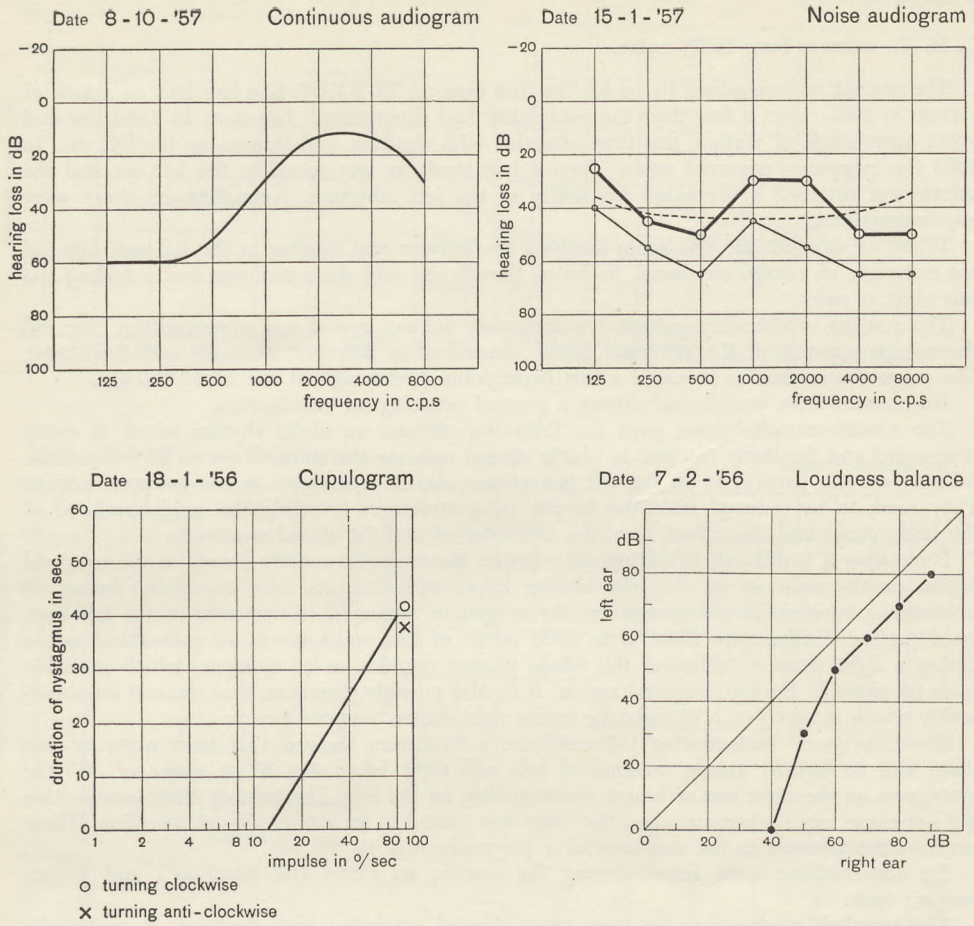


Figure 46

The threshold audiograms for pure tones showed a bass deafness in the right ear which varied considerably. Subjectively too the hearing varied considerably. A continuous threshold audiogram was also made. Fowler's test (binaural balance) showed the presence of a moderate recruitment in the right ear at 250 c.p.s. Langenbeck's audiogram was of the ganglion type. The tones had the same pitch in both ears; they sounded impure, however, in the right ear. In the course of two years the audiogram showed a transition to type II (figure 46).

Case 5

C. H.-V., woman, born 1920, nurse.

The patient was examined by us for the first time on 12-2-1957. She had had an attack of vertigo in 1952; after a few days the complaints had disappeared, however. In 1954 she had again complained of vertigo, this time attended with deafness and tinnitus in the left ear. In 1955 the symptoms occurred again. Then a bass deafness was found in the left ear and the caloric test revealed a decreased irritability of the left labyrinth. Neurologically there were no abnormalities.

When we saw her she was again troubled by deafness and tinnitus in the left ear. She did not complain of vertigo any more, however, though she said she sometimes had a feeling she was sinking away.

The routine otorhinolaryngological examination did not reveal any abnormalities, nor did the roentgenograms of the temporal bones (according to Schüller, Stenvers and Steenhuis). The internal examination revealed a low hemoglobin percentage of the blood (60 %).

No abnormalities were found during a general neurological examination.

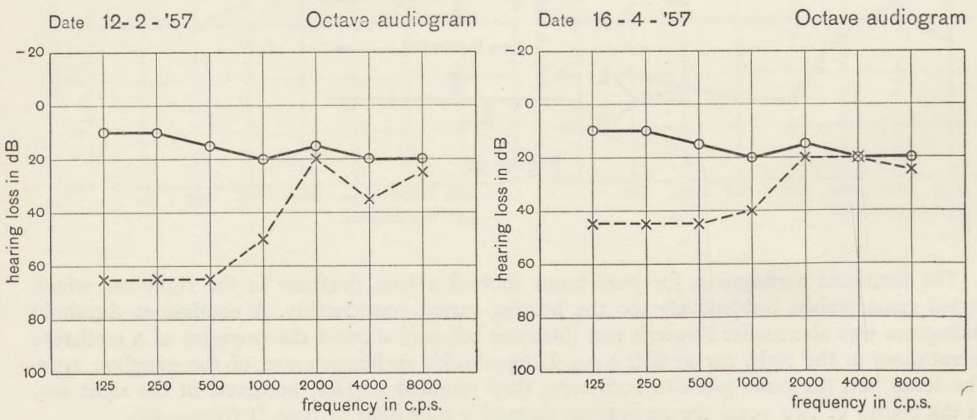
The electro-encephalogram gave the following picture: an alpha rhythm which is easily suppressed and the lively reaction to photic stimuli indicate the strained nerves of the patient. Striking are the paroxysms on the left (sometimes also on the right) in the temporal region. They seem to be induced from the deeper lying structures (possibly the neighbourhood of the brain stem) and sometimes have the character of a slight stimulus activity.

Sometimes it is difficult to distinguish whether the strongest activity occurs in the temporal region on the right or on the left. During hyperventilation and also especially during an intravenous injection of chlorpromazine the symptoms appeared to dominate in the left temporal region. Furthermore there were little series of fast spike waves of subcortical origin during a light sleep. Conclusion: the whole picture reminds us of epilepsy, which probably finds its origin in the left temporal region. It is, also possible, however, that there is an abnormality which is based on a disturbance in the right centro-temporal region.

The calorigram made during the vestibular examination showed that with water of 44° there was an equally strong reaction of left and right labyrinths. With water of 30° the nystagmus on the right was of longer duration than on the left. The turning chair examination did not cause any nystagmus when the chair was turned in an anti-clockwise direction. These results point perhaps to the existence of a preponderance to the left.

No abnormalities were found during the marche en étoile and Romberg's and Unterberger's tests.

The threshold audiograms for pure tones showed a varying bass deafness. A continuous threshold audiogram was also made. Fowler's test (binaural balance) showed the presence of recruitment in the left ear at 500 c.p.s. The Langenbeck's noise audiogram was of the ganglion type. The tones had the same pitch in both ears but sounded impure in the left ear. In the speech audiogram was a hearing loss of about 40 dB, while speech perception was 90 % when sufficiently amplified (figure 47).



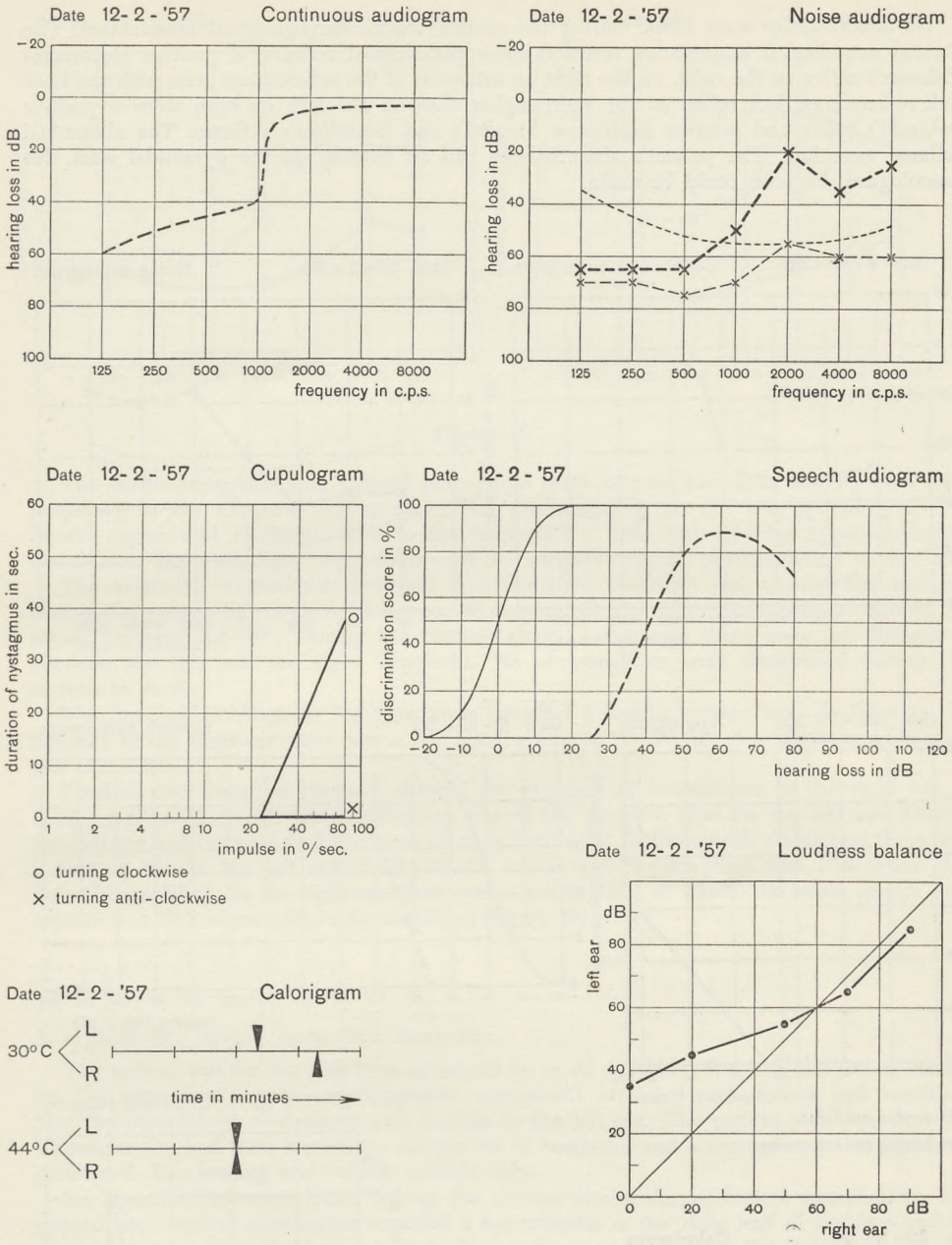


Figure 47

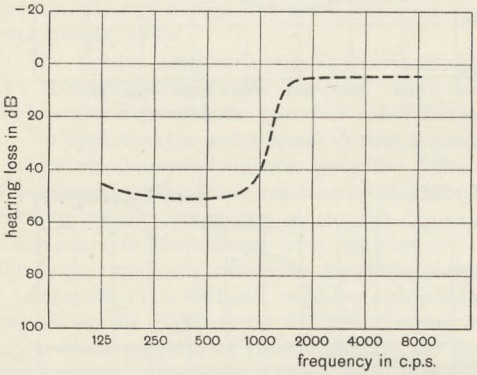
Case 6

E. A.-S., woman, born 1915, housewife.

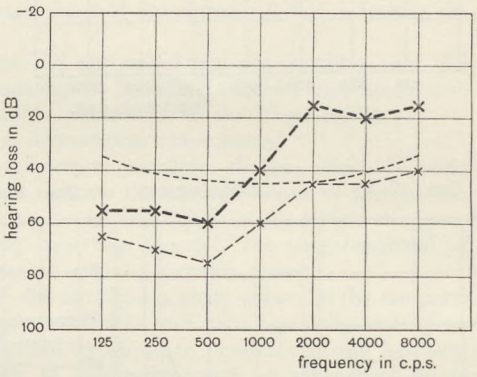
The patient was seen by us for the first time on 29-5-1956. She had had an otitis media in 1922 and she said her hearing had gradually become worse since that time. She had 2 children and during pregnancy her deafness had increased, she said. No deafness occurred in the family. She had never been ill. Since a short time she had been troubled by tinnitus in the left ear. She did not complain of vertigo. The hearing was not varying she said.

No abnormalities were found during the routine otorhinolaryngological examination. The general neurological examination revealed some pathological reflexes: a positive Hoffmann-Trömner's reflex on the right, on the right an extension of the reflexogenic zone with the knee jerk reflex, knee jerk reflex on the right higher than on the left, on both sides a positive Babinski's reflex and positive Bechterew Mendel's and Rossolimo's reflexes. The abdominal reflexes were low. The patient's abnormalities had all bearing on the pyramidal tract. No neurological diagnosis could be made.

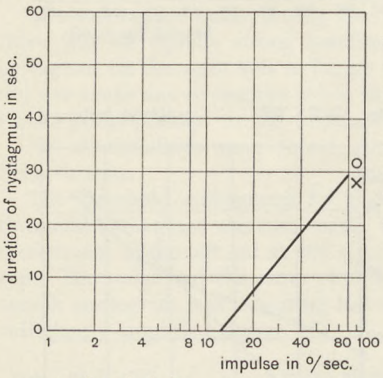
Date 9 - 10 - '56 Continuous audiogram



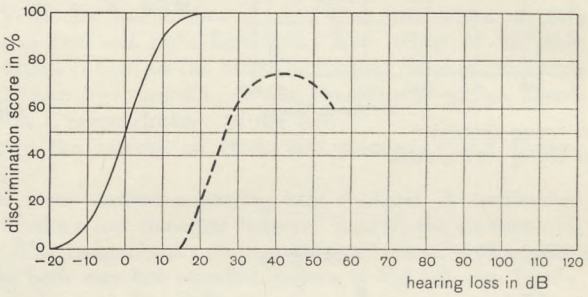
Date 29 - 5 - '56 Noise audiogram



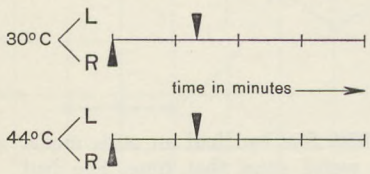
Date 30 - 5 - '56 Cupulogram



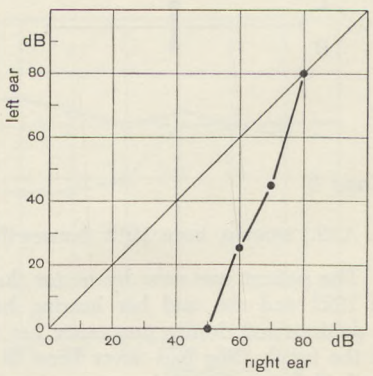
Date 9 - 10 - '56 Speech audiogram



Date 30 - 5 - '56 Calorigram



Date 29 - 5 - '56 Loudness balance



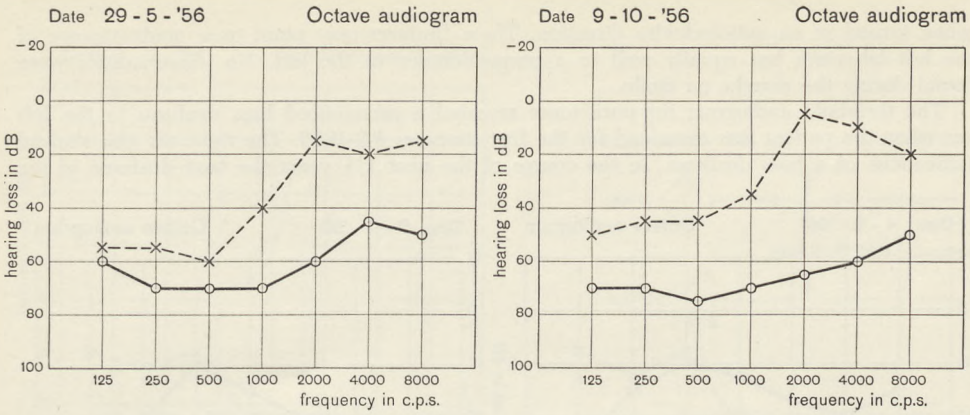


Figure 48

The electro-encephalogram showed paroxysmal theta activity and delta activity of a high frequency in the temporal region, sometimes predominantly on the left, spreading to the frontal region and sometimes also to the back and a little activated by hyperventilation. Conclusion: there are light abnormalities in the temporal region, predominantly on the left.

The vestibular examination revealed that the right labyrinth was clearly less irritable during the caloric test than the left one. Nystagmus on the right could only be evoked by means of water of 20°. During the turning chair examination there were no differences between the left and the right labyrinths. No abnormalities were discovered during the marche en étoile.

The threshold audiograms for pure tones revealed a hardly varying bass deafness in the left ear; in the right ear there was a flat hearing loss of 60–70 dB. A continuous audiogram was also made.

Fowler's test (binaural balance) showed the presence of recruitment in the right ear at 2000 c.p.s. Langenbeck's noise audiogram was of the ganglion type for the left ear. During the balance test the tone had the same pitch in both ears. In the speech audiogram there was a hearing loss in the left ear of 25–30 dB, while speech perception was 75 % when sufficiently amplified. In the right ear there was a hearing loss of 45–50 dB while speech perception was 70 % when sufficiently amplified (figure 48).

Case 7

A. v. d. H.-W., woman, born 1912, housewife.

The patient was for the first time examined by us on 4-9-1957. She had had fits of vertigo for 1½ years (not a pronounced rotatory movement), attended with nausea and vomiting. She also complained of deafness and tinnitus in the left ear. The patient was very nervous. Elsewhere she had been treated for complaints of headache and a septum resection had been performed. The hearing was varying considerably.

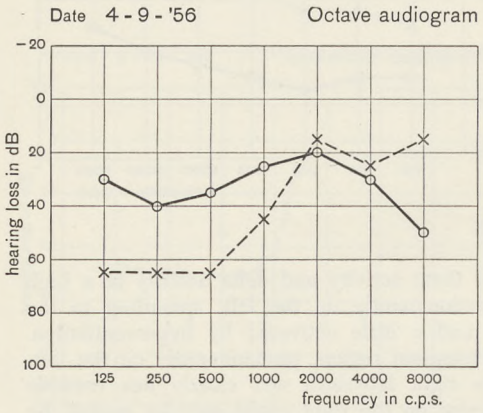
No abnormalities were found during the routine otorhinolaryngological examination. The general neurological examination revealed a hyperalgesia in the right half of the face and all over the right half of the body. A slight hyperreflexia was found in the right arm and leg. The abdominal reflexes too were a little higher on the right than on the left. There were no pathological reflexes and no disturbances in coordination. The hyperpathia on the right indicated an affection of the vegetative nervous system. The electro-encephalogram showed a slight diffuse beta activity. Response to photic stimuli, more on the left than on the right and some harmonic and subharmonic response.

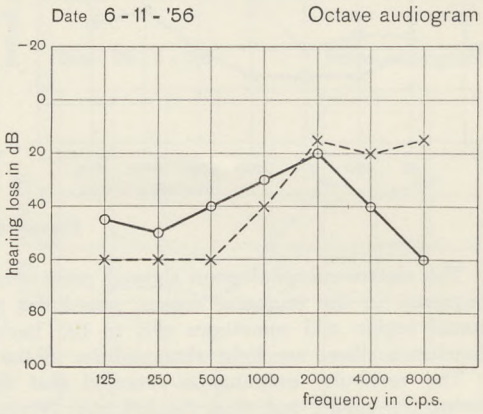
Conclusion: the asymmetrical flash reaction is a little abnormal. For the rest the picture did not show any abnormalities.

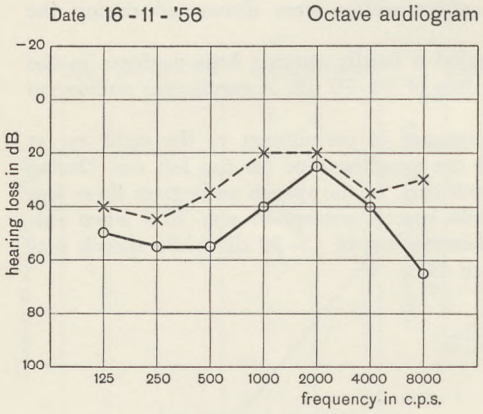
When syringed with water of 30° there was an equal irritability of both labyrinths; with water of 44° no nystagmus on the right could be evoked. The turning chair examination revealed that the nystagmus was of longer duration when turned in a clockwise direction than

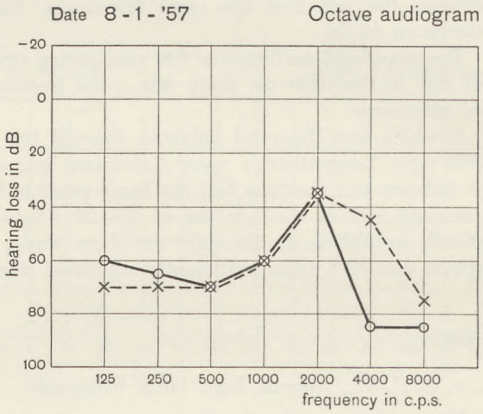
when turned in an anticlockwise direction. These findings may point to a predominance of the left labyrinth but equally well to a preponderance to the left. No abnormalities were found during the marche en étoile.

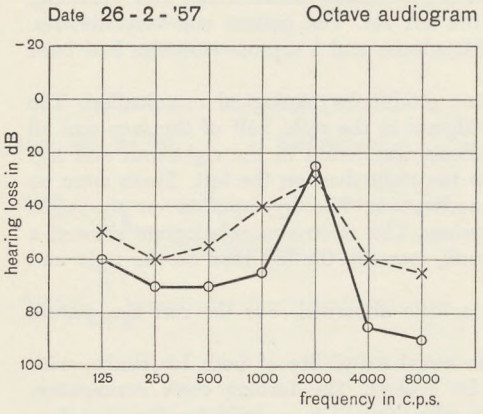
The threshold audiogram for pure tones revealed a pronounced bass deafness in the left ear when the patient was examined for the first time, on 4-9-1956. The right ear also showed indications of a bass deafness. In the course of the next 1½ years the bass deafness in the

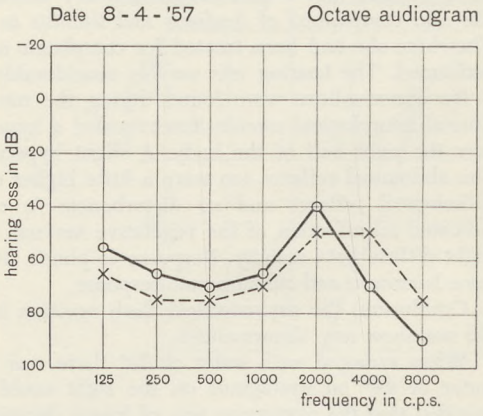












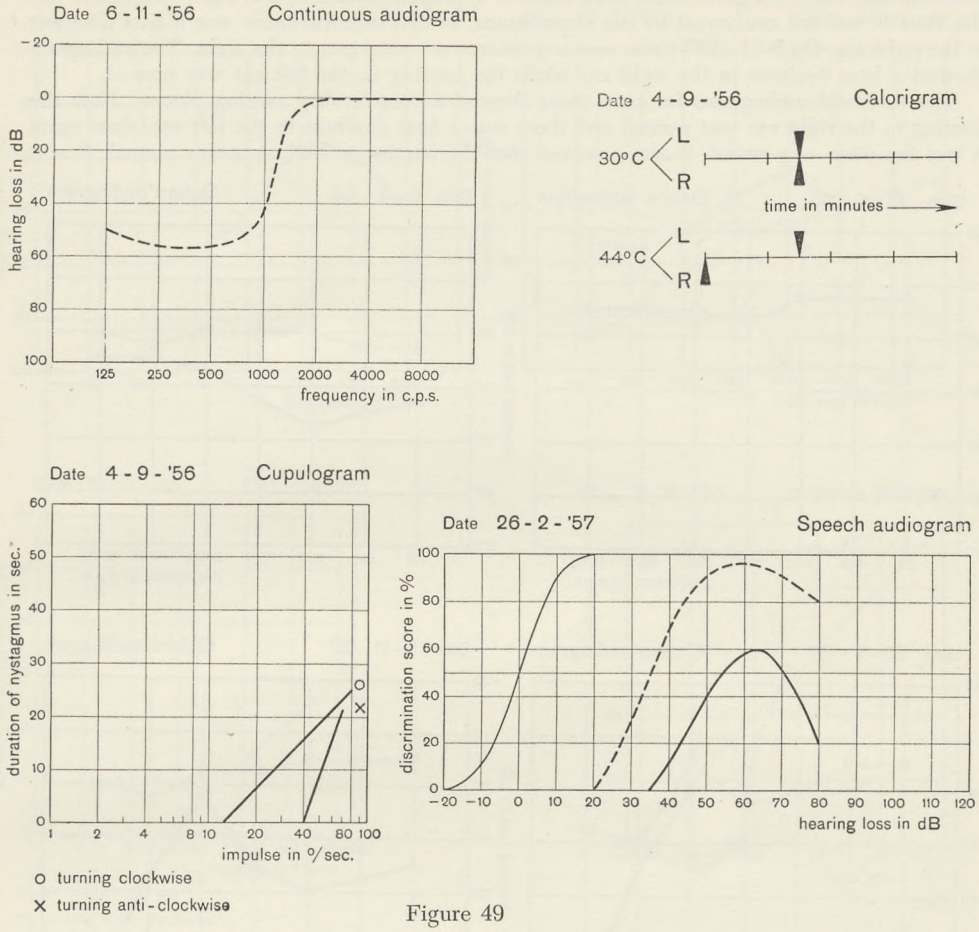


Figure 49

left ear varied a little but this was especially pronounced in the right ear. During this period there was a clear transition to type II.

A few times a continuous audiogram was made. On 26-2-1957 a speech audiogram was made, showing a hearing loss in the left ear of 35 dB, while speech perception was 100 % when sufficiently amplified. In the right ear there was a hearing loss of 60 dB, speech perception being 60 % when sufficiently amplified (figure 49).

Case 8

P. v. V., man, born 1914, plumber.

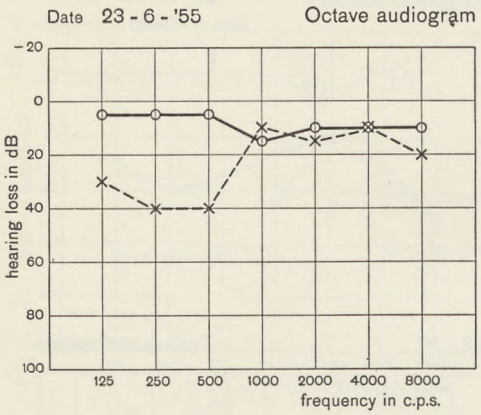
The patient was examined by us for the first time on 23-6-1955. Nine weeks before he had had a touch of influenza. Since that time he had been complaining of fits of rotatory vertigo attended with nausea and vomiting. The external world turned to the left. There were no hearing complaints but there was a slight tinnitus.

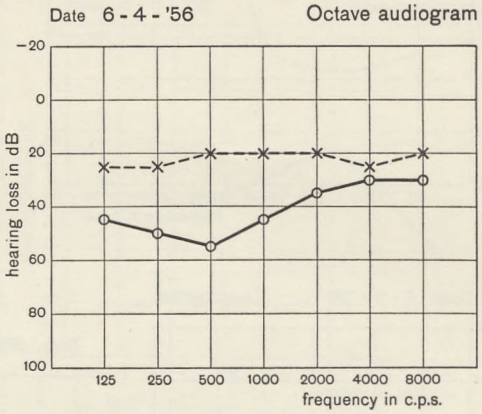
No abnormalities were found during the routine otorhinolaryngological examination, nor in the roentgenograms of skull and temporal bones. During the general neurological examination no abnormalities could be found either.

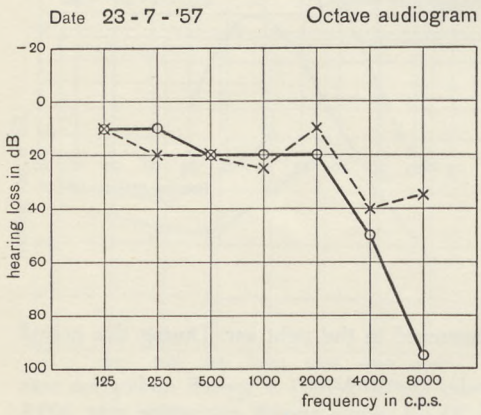
During the first vestibular examination on 9-8-1955 the calorigram showed a preponderance to the right; this was confirmed by the cupulogram. The marche en étoile revealed a deviation to the left. At the time the hearing was normal on both sides. A second vestibular examination

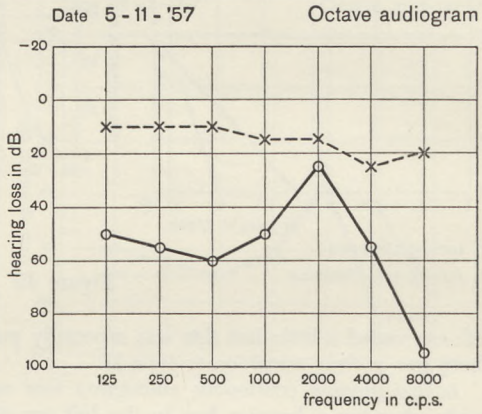
revealed that the calorigram once more showed a preponderance to the right (6-6-1956) but this time it was not confirmed by the cupulogram; at that moment there was a bass deafness in the right ear. On 5-11-1957 there was a spontaneous nystagmus to the right. The audiogram showed a bass deafness in the right ear while the hearing in the left ear was normal.

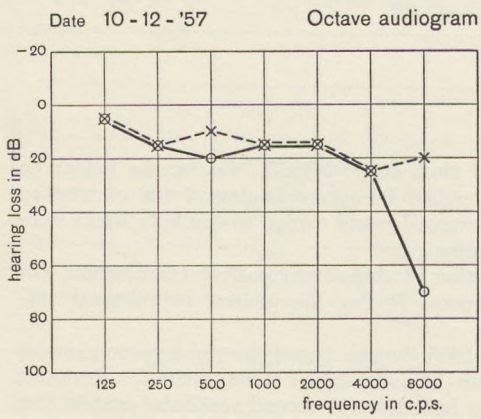
The threshold audiograms for pure tones showed a considerably varying picture. Now the hearing in the right ear was normal and there was a bass deafness in the left ear, then again it was the other way round. It also occurred that the hearing in both ears was normal. It was

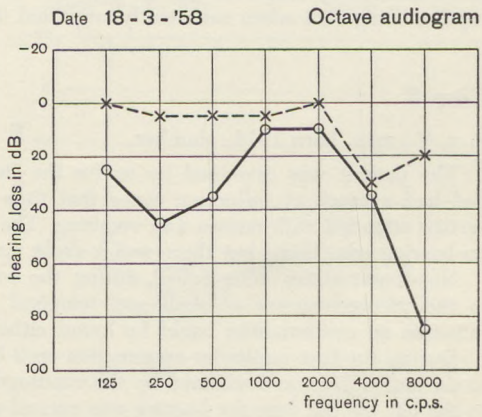












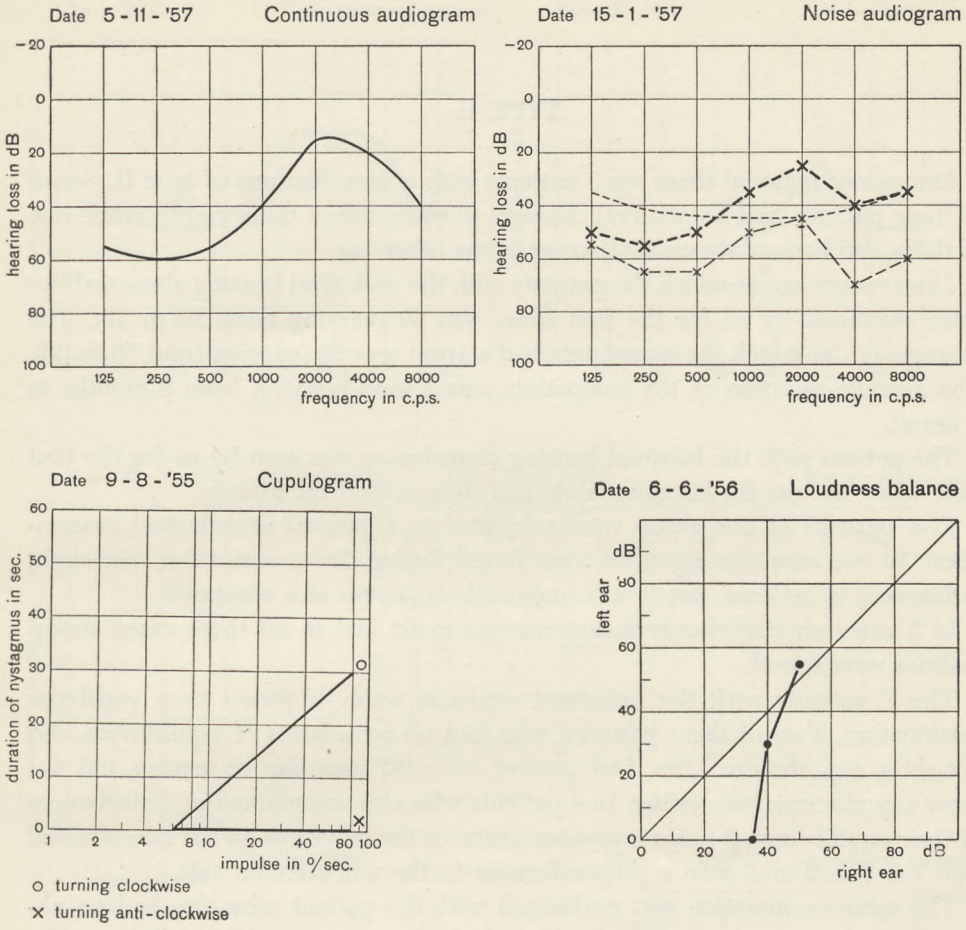


Figure 50

found that there was a transition to type II for the right ear. Fowler's test on 6-6-1956 (binaural balance) showed the presence of recruitment in the right ear at 500 c.p.s. The tone had a higher pitch in the right than in the left ear (diplacusis). Langenbeck's noise audiogram was of the ganglion type. A continuous audiogram was also made (figure 50).

TYPE II

Among our material there are 8 patients with a bass deafness of type II. Seven of these patients had a unilateral deafness (3 times left, 4 times right); while one of them also had an abnormal hearing in the other ear.

The average age at which the patients with the unilateral hearing abnormalities were examined by us for the first time, was 30 (varying from 22 to 37). The average age at which the complaints had started was 29 (varying from 20 to 36). The average duration of the complaints was 1 year (varying from 2 months to 2 years).

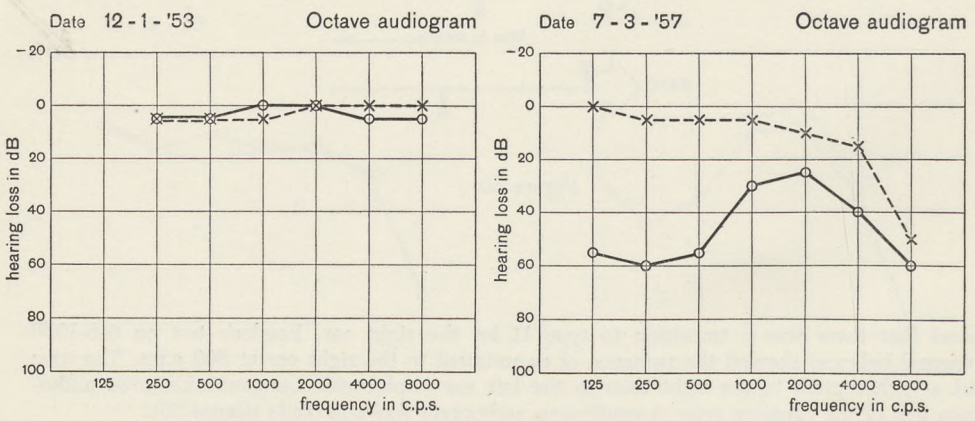
The patient with the bilateral hearing disturbance was seen by us for the first time when he was 26; his complaints had then existed for 2 years.

Five patients of this group were subjected to a general neurological examination. In one case abnormalities were found during this examination, viz. slight differences in reflexes, but it was impossible to arrive at a diagnosis.

In 5 cases an electro-encephalogram was made and in all these cases abnormalities were found.

The 7 patients with the unilateral deafness were subjected to a vestibular examination. Two of these patients, who had no complaints of equilibrium, did not show any abnormalities. One patient who did complain of vertigo, did not show any abnormalities either. In 4 patients who also complained of disturbances in their equilibrium, 3 times a predominance of the labyrinth on the non-affected side was found and once a preponderance to the non-affected side.

The same examination was performed with the patient who also had an abnormal hearing on the other side. His labyrinths proved to be hardly irritable. Cold water (20°) could evoke a trace of nystagmus on the side of the bass deafness (predominance of the labyrinth on the side of the bass deafness?).



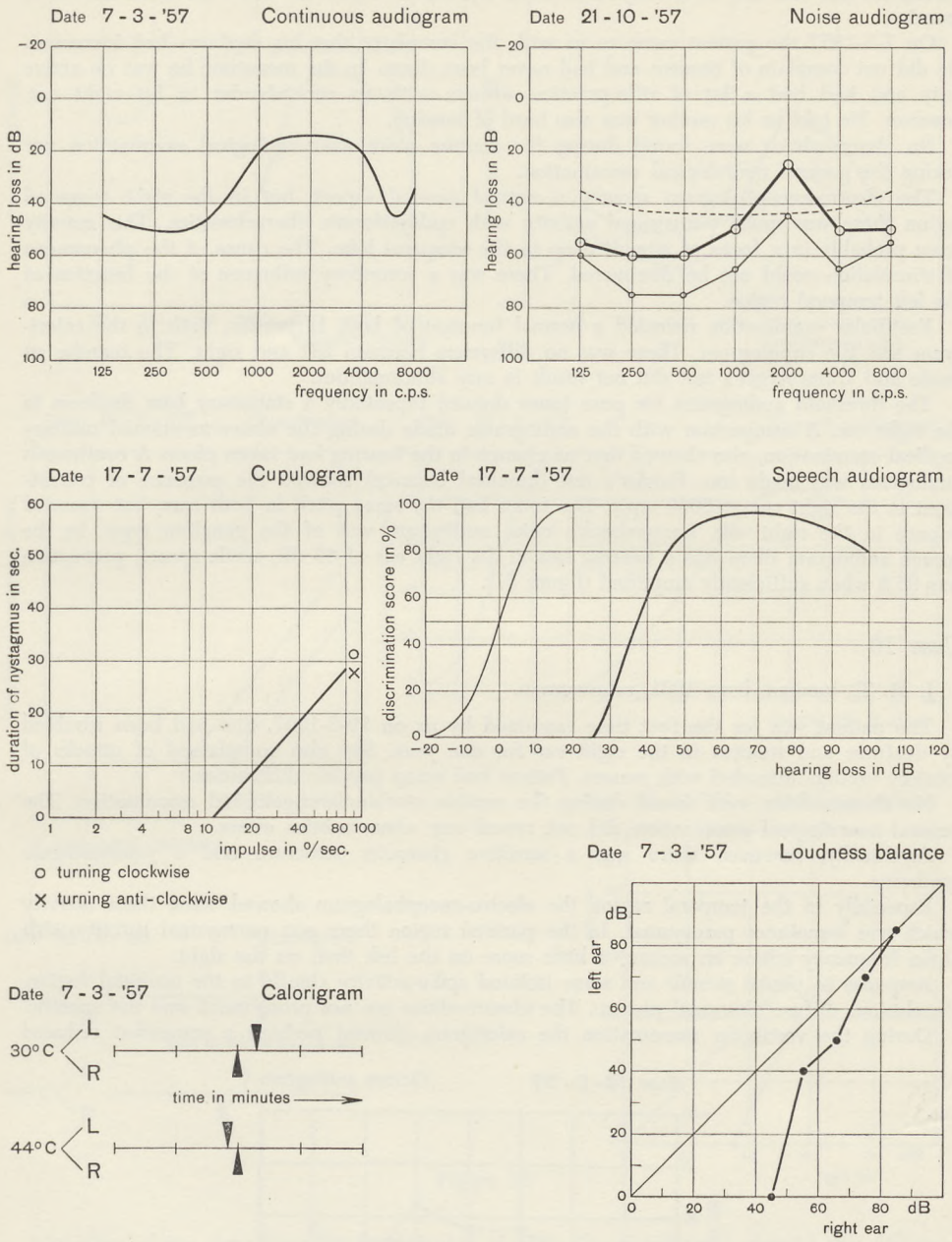


Figure 51

Case 9

D. A. In 't V., born 1935, undergraduate.

The patient was seen by us for the first time on 7-3-1957. In 1953 he had been medically examined for civil aviation. No hearing abnormalities were found then (audiogram). In 1956 he had been medically examined for the military airforce. On that occasion a bass deafness in the right ear was accidentally found, characterized by the presence of recruitment. The

calorigram showed that both labyrinths were equally irritable. No other abnormalities were found.

On 7-3-1957 the patient came to us with the complaint that his deafness had increased. He did not complain of tinnitus and had never been dizzy. In the meantime he was on active duty and had had a lot of rifle-practice, always with an ear-defender in his right ear, however. He told us his mother was also hard of hearing.

No abnormalities were found during the routine otorhinolaryngological examination nor during the general neurological examination.

The electro-encephalogram showed a normal general aspect, but in the right temporal region there was much paroxysmal activity with epileptiformic characteristics. This activity arose probably in a focus of stimuli deep in the temporal lobe. The cause of the phenomena of stimulation could not be discovered. There was a secondary influence of the function of the left temporal region.

Vestibular examination revealed a normal function of both labyrinths, both in the calorigram and the cupulogram. There was no difference between left and right. The marche en étoile and Unterberger's test did not result in any abnormalities.

The threshold audiograms for pure tones showed repeatedly a stationary bass deafness in the right ear. A comparison with the audiograms made during the above-mentioned military medical examination, also showed that no change in the hearing had taken place. A continuous audiogram was made too. Fowler's test (binaural balance) showed the presence of recruitment in the right ear at 5000 c.p.s. The tones had the same pitch in both ears, but sounded impure in the right ear. Langenbeck's noise audiogram was of the ganglion type. In the speech audiogram there was a hearing loss in the right ear of 35 dB, while speech perception was 95 % when sufficiently amplified (figure 51).

Case 10

J. M. T., woman, born 1921, saleswoman.

The patient was for the first time examined by us on 19-3-1957. She had been troubled by deafness and tinnitus in the right ear for one year. She also complained of attacks of rotatory vertigo, attended with nausea. Patient had many psychic difficulties.

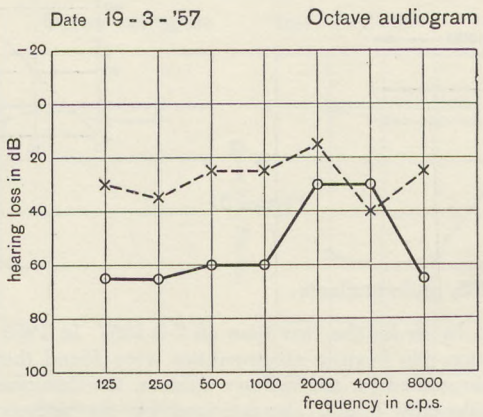
No abnormalities were found during the routine otorhinolaryngological examination. The general neurological examination did not reveal any abnormalities either.

Psychically, however, there was a sensitive character structure and a psychasthenic syndrome.

Especially in the temporal region the electro-encephalogram showed some theta activity which was sometimes paroxysmal. In the parietal region there was paroxysmal activity with alpha frequency (ritme en arceau) a little more on the left than on the right.

Response to photic stimuli and some isolated spike-activity chiefly in the occipital region. Conclusion: diffuse abnormal picture. The abnormalities are not pronounced and not specific.

During the vestibular examination the calorigram showed perhaps a somewhat reduced



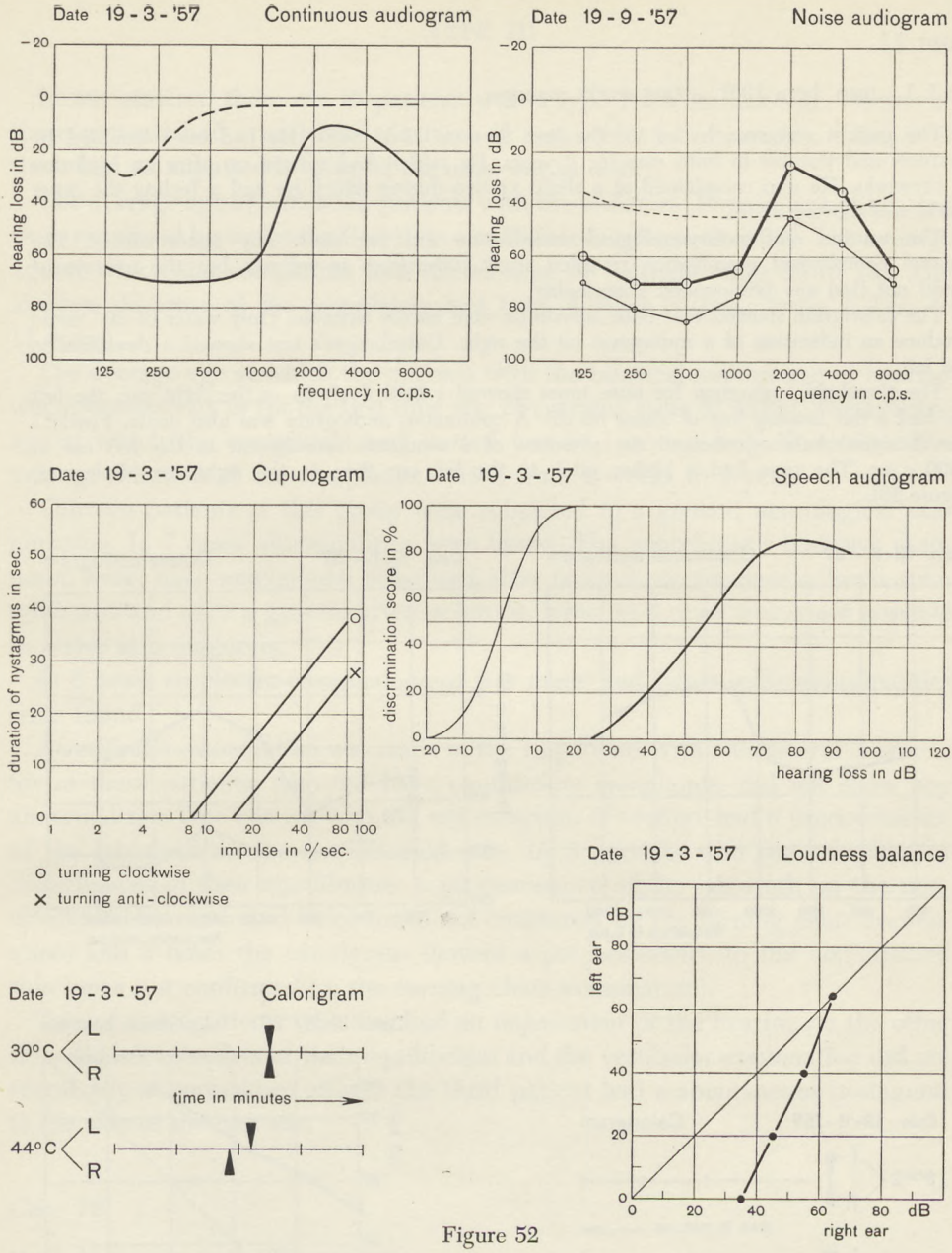


Figure 52

irritability of the right labyrinth (with water of 44°). The cupulogram showed this difference even more pronounced. This might point to a predominance of the left labyrinth. A repetition of the caloric test half a year later gave the same results. The marche en étoile and Unterberger's test showed a deviation to the left. Romberg's test did not reveal any abnormalities.

The threshold audiograms for pure tones repeatedly showed an almost unvarying bass deafness in the right ear. A continuous audiogram was also made. Fowler's test (binaural balance) showed the presence of recruitment in the right ear at 250 c.p.s. The tones had the same pitch in both ears but sounded impure in the right ear. Langenbeck's noise audiogram was of the ganglion type. In the speech audiogram there was a hearing loss of about 50 dB in the right ear, while speech perception was 85 % when sufficiently amplified (figure 52).

Case 11

H. J. L., man, born 1931, acting works manager.

The patient was seen by us for the first time on 12-11-1957. He had been troubled by deafness and tinnitus in both ears for 2 years. He said it had sprung up after an explosion of fireworks. He also complained of a slight vertigo during which he had a feeling the outer world was moving about.

The routine otorhinolaryngological examination did not show any abnormalities. The general neurological examination revealed slight differences in reflexes but the neurologist could not find any pronounced abnormality.

The calorigram showed that both labyrinths were hardly irritable. Only water of 20° could produce an indication of a nystagmus on the right. Unterberger's test showed a deviation to the left.

The threshold audiogram for pure tones showed a bass deafness in the right ear; the left ear had a flat hearing loss of about 80 dB. A continuous audiogram was also made. Fowler's test (binaural balance) showed the presence of a moderate recruitment in the left ear at 2000 c.p.s. The tone had a higher pitch in the left ear than in the right ear (diplacusis) (figure 53).

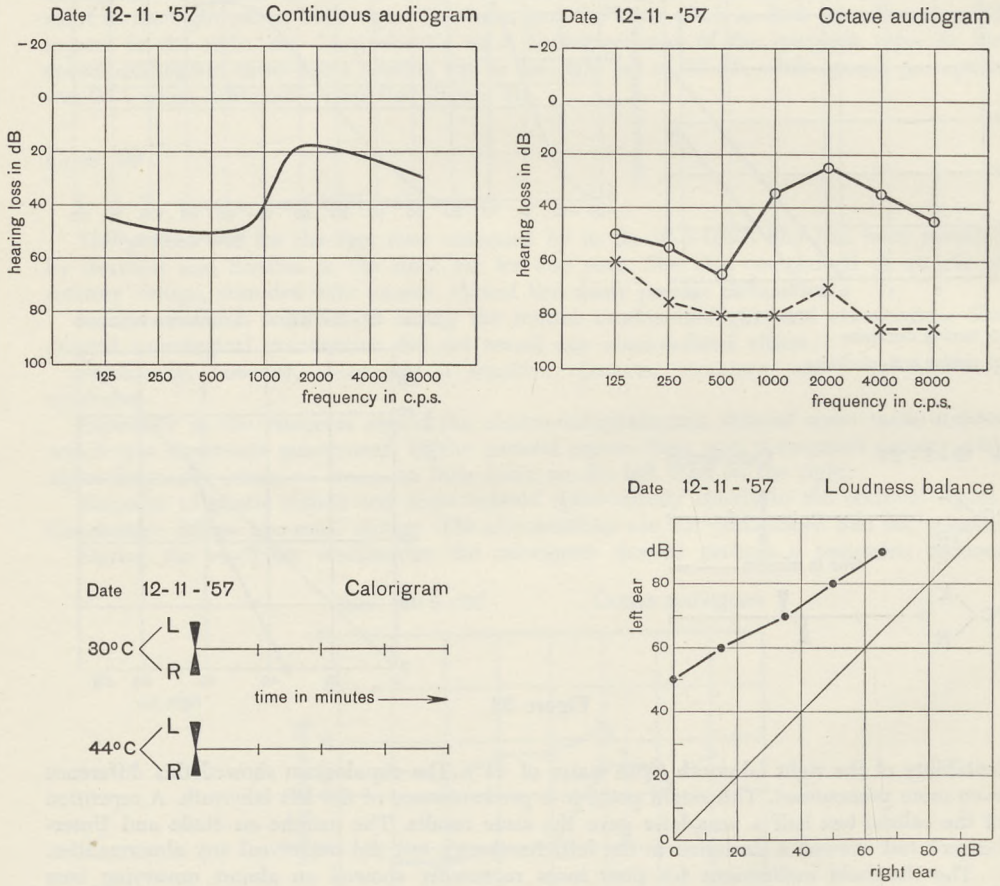


Figure 53

TYPE III

In our material there are 15 patients with a type III bass deafness. Twelve of these patients had a unilateral deafness (4 times left, 8 times right), while 3 of them had an impaired hearing in the other ear as well.

The average age at which the patients with the unilateral impairment of hearing were examined by us for the first time was 58 (varying from 46 to 73). The average age at which the complaints had started was 54 (varying from 35 to 73). The average duration of the complaints had been 4 years (varying from 2 days to 11 years).

The average age at which the patients with the bilateral impairment of hearing were examined by us for the first time was 58 (varying from 57 to 61). The average age at which the complaints had started was 58 (varying from 57 to 60). The average duration of the complaints varied from 2 weeks to 1 year.

Thirteen patients of this group were subjected to a general neurological examination. In 7 cases abnormalities were found. The neurologist's tentative diagnoses were: once arachnoidal adhesions after trauma capitis, once a brain stem syndrome and once a general arteriosclerosis, while in 4 cases it was not possible to arrive at a diagnosis.

In 3 cases an electro-encephalogram was made and twice some abnormalities were found.

A vestibular examination was made of the 12 patients with a unilateral deafness. Six of these patients, who did have equilibrium complaints, did not show any abnormalities. One patient who did not complain of vertigo, had a predominance of the labyrinth on the non-affected side. In 5 patients who did complain of disturbances in their equilibrium, a predominance of the labyrinth on the non-affected side was found twice (once not confirmed by the turning chair examination) and 3 times the calorigram showed a preponderance to the non-affected side (once not confirmed by the turning chair examination).

Two of the 3 patients who also had an impairment of the hearing on the other side, did not complain of their equilibrium and the vestibular examination did not reveal any abnormalities either. The third patient had a spontaneous nystagmus to the side of the best ear.

Case 12

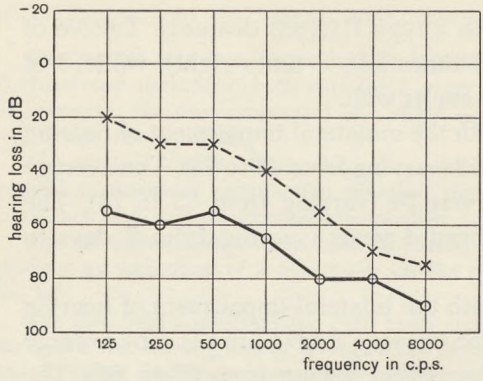
N. C. de L., man, born 1898, pensioner.

The patient was for the first time examined by us on 13-9-1955. He had suffered from deafness and tinnitus in the right ear for 6 weeks. He did not complain of vertigo. He did complain of a stopped-up nose and for this he was treated (removal of polypus and puncture of the maxilla).

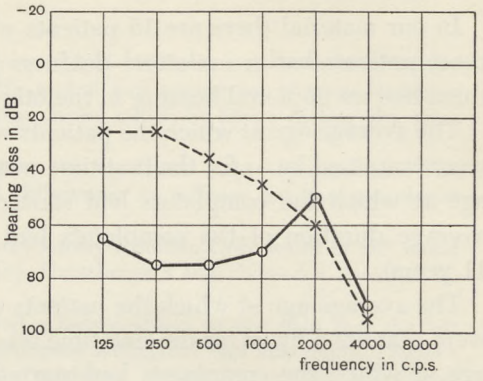
The routine otorhinolaryngological examination did not reveal any abnormalities in the ear drums, nor were any abnormalities found in the roentgenograms of the temporal bones.

The first general neurological examination revealed subjectively a difference in irritability of the cornea to the detriment of the right one. Objectively no difference could be proved. There was a total hypalgesia and hypesthesia on the right side which was taken to be

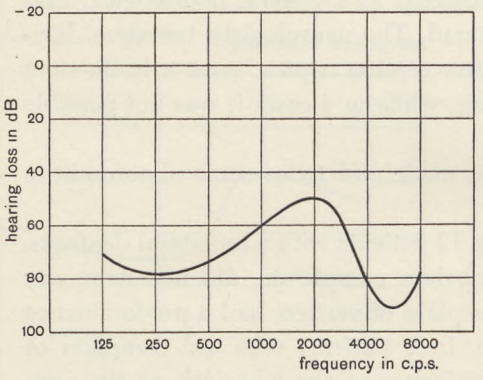
Date 13 - 9 - '55 Octave audiogram



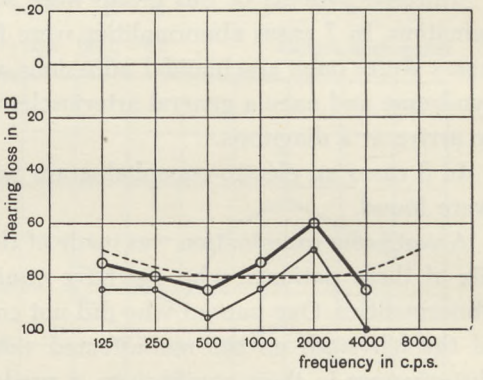
Date 27 - 8 - '56 Octave audiogram



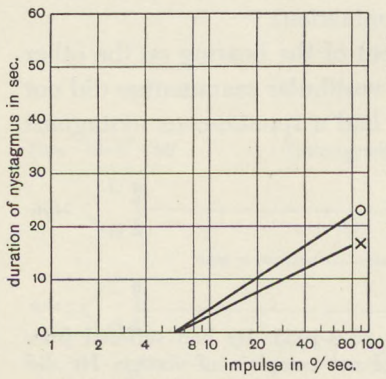
Date 20 - 11 - '56 Continuous audiogram



Date 21 - 1 - '57 Noise audiogram



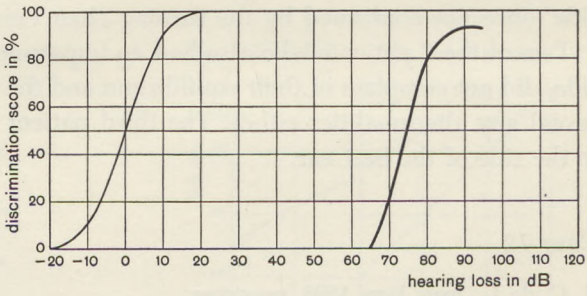
Date 27 - 8 - '54 Cupulogram



o turning clockwise

× turning anti-clockwise

Date 25 - 2 - '57 Speech audiogram



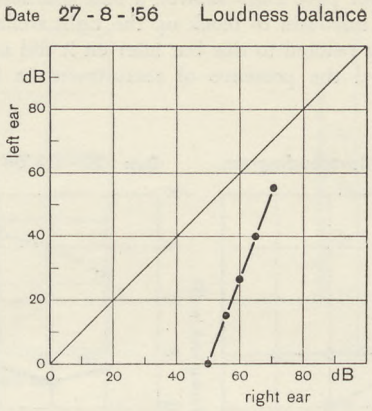


Figure 54

psychogenic. When this examination was repeated there were dubious abnormalities in the glossopharyngeal nerve and in the reflexes of the left arm, which combined with the audiological and vestibular findings might point in the direction of a brain stem syndrome. A renewed check revealed that the abnormalities varied considerably and the neurologist considered them to be psychogenic.

An ophthalmic examination revealed a stage III arteriolosclerosis in fundo.

The vestibular examination showed a predominance of the left labyrinth both in the calorigram and the cupulogram. During the marche en étoile there was a deviation to the right.

The threshold audiograms for pure tones showed a constant bass deafness in the right ear, except during the first examination. A continuous audiogram was made as well. Fowler's test (binaural balance) showed the presence of recruitment in the right ear at 250 c.p.s. The tones in both ears had the same pitch but were impure in the right ear. Langenbeck's noise audiogram was of the ganglion type. In the speech audiogram there was a hearing loss in the left ear of 50 dB, while speech perception was 100 % when sufficiently amplified. In the right ear there was a hearing loss of 75 dB while speech perception was 95 % when sufficiently amplified (figure 54).

Case 13

A. de V., man, born 1886, pensioner.

The patient was seen by us for the first time on 23-1-1950. He was troubled by tinnitus which had come about after a cold. The audiogram then showed a presbycusis. No abnormalities were shown by the marche en étoile and Romberg's test.

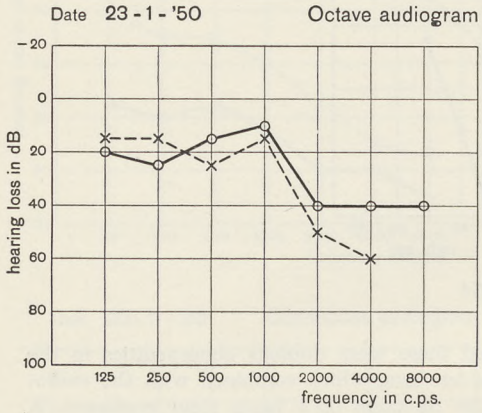
The patient came to us again on 20-11-1954. He had been troubled by tinnitus in the right ear and deafness for 2 days. He had also been slightly dizzy (no rotatory vertigo). The complaints had cropped up without any apparent cause.

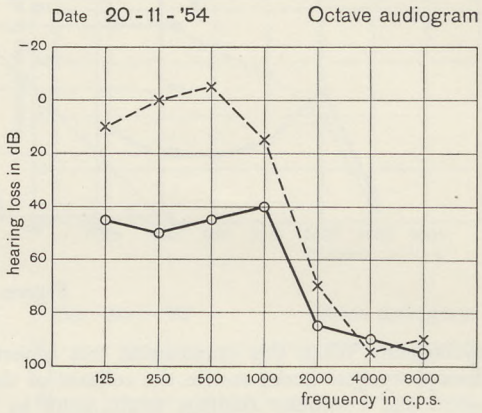
The routine otorhinolaryngological examination and the roentgenograms of skull and temporal bones did not show any abnormalities.

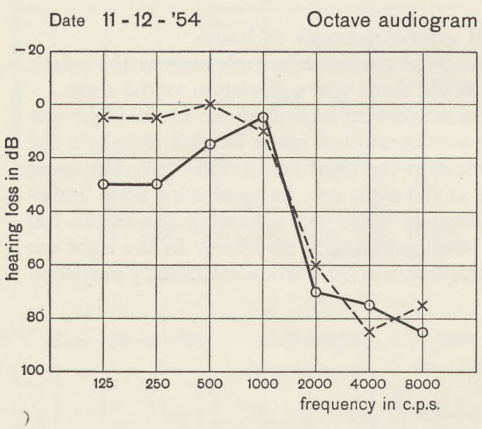
A general neurological examination revealed very low to non-existent Achilles tendon reflexes on both sides. Footsoles indifferent. The vibratory sensibility in legs and cristae was reduced. It was considered likely that this was a case of cerebral arteriosclerosis.

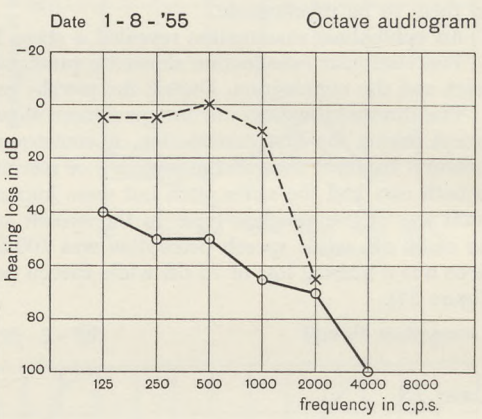
A first vestibular examination on 9-3-1956 showed a normal function of both labyrinths, both in calorigram and cupulogram. There was no difference between left and right. No abnormalities were found during the marche en étoile. During a new caloric test on 24-12-1956 it seemed as if there was a preponderance to the left. The marche en étoile and Unterberger's test revealed a deviation to the right. During a third caloric test on 1-5-1957 there was again no pronounced difference between the two labyrinths. The marche en étoile then showed a deviation to the left.

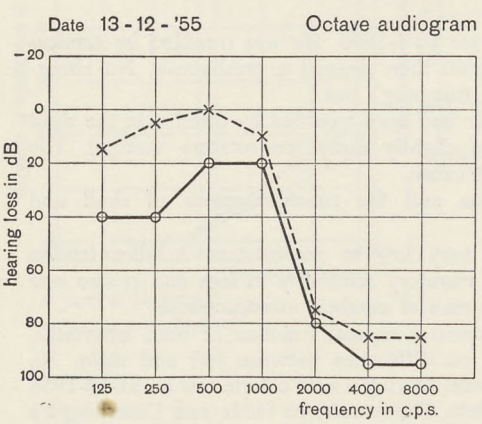
The threshold audiograms for pure tones showed a bass deafness which varied considerably. The patient was treated with novocain to block up the right stellate ganglion. In the beginning it seemed as if the hearing reacted to this but later on it did not do so any more. Fowler's test (binaural balance) showed the presence of recruitment in the right ear at 250 c.p.s. (figure 55).

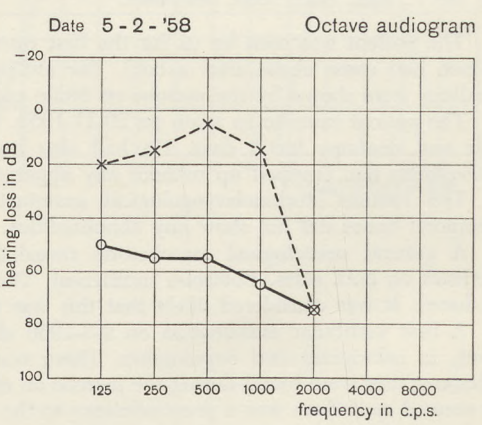












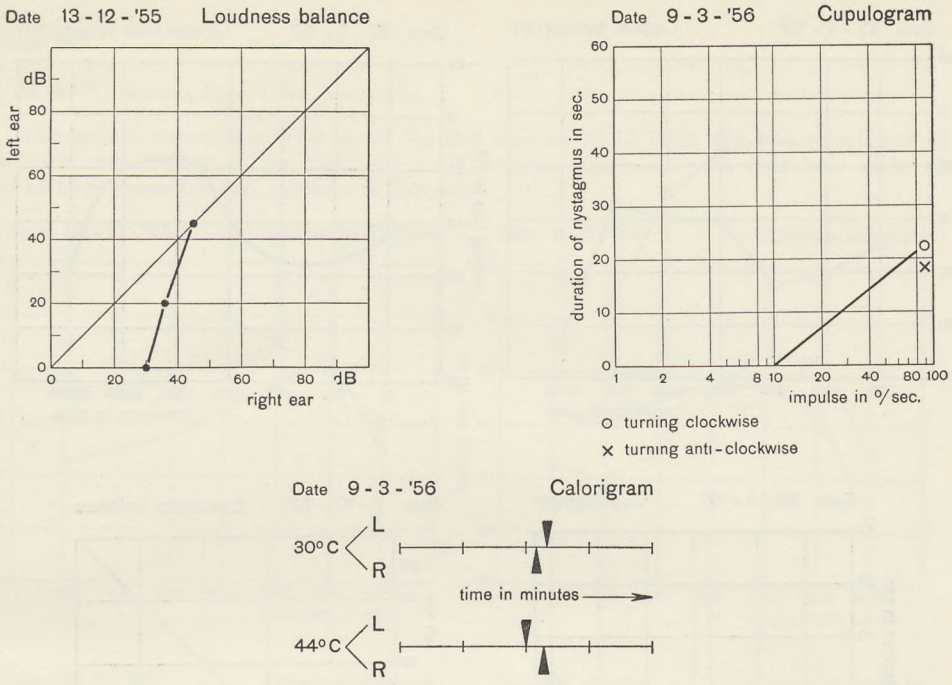


Figure 55

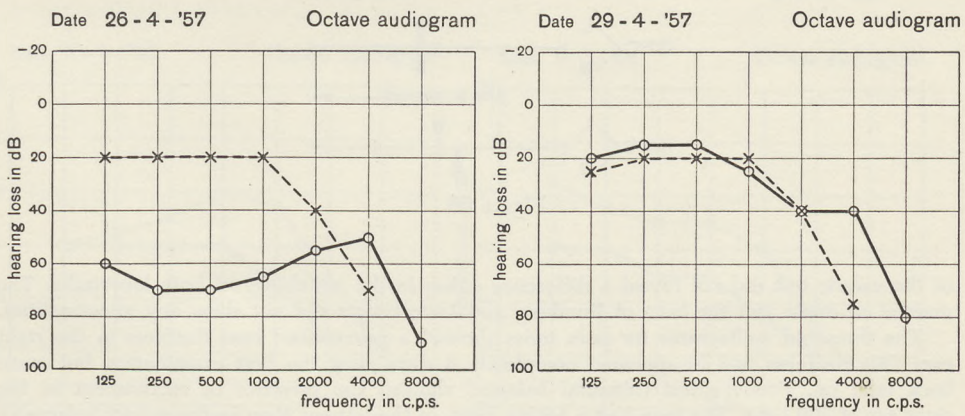
Case 14

J. C. de Z., woman, born 1910, tailor's cutter.

The patient was seen by us for the first time on 26-4-1957. She had suffered from deafness and tinnitus in the right ear. The hearing varied considerably, she also had periods that is was very good. She also had fits of vertigo, attended with nausea and vomiting.

The routine otorhinolaryngological examination and the general neurological examination did not show any abnormalities.

The vestibular examination showed a normal function of both labyrinths, both in the calorigram and the cupulogram. There was no difference between left and right. Repetition



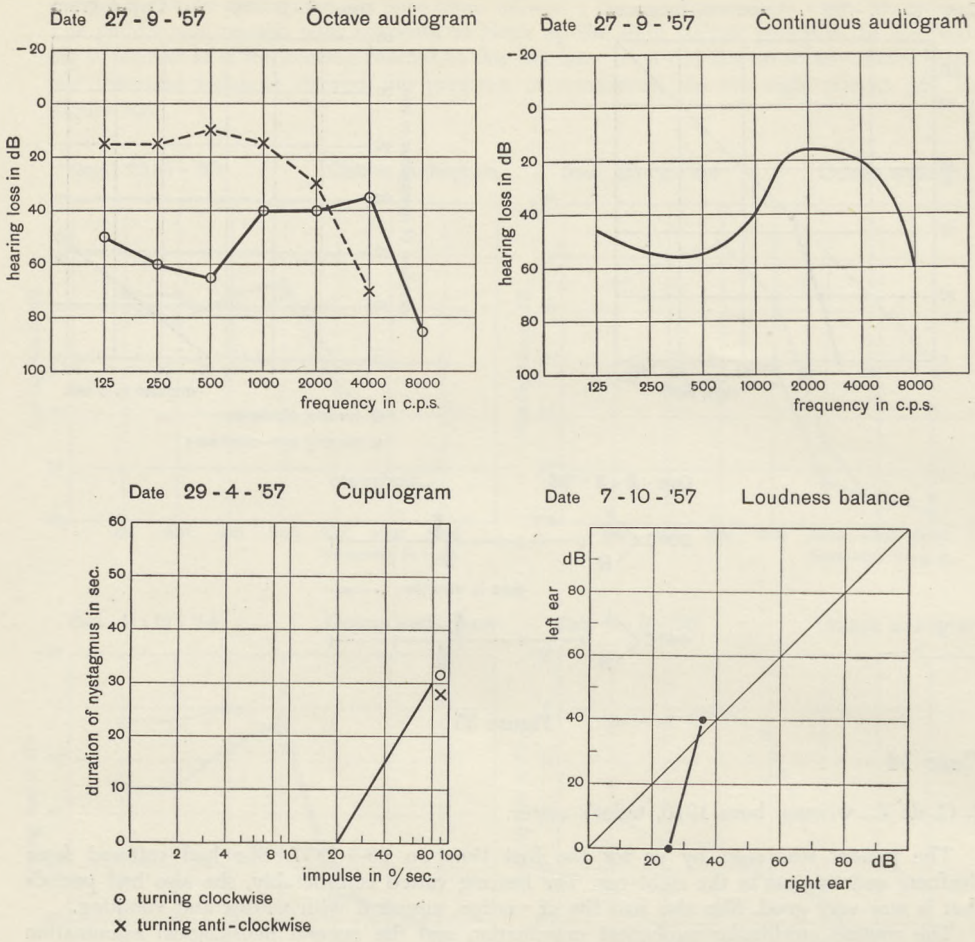


Figure 56

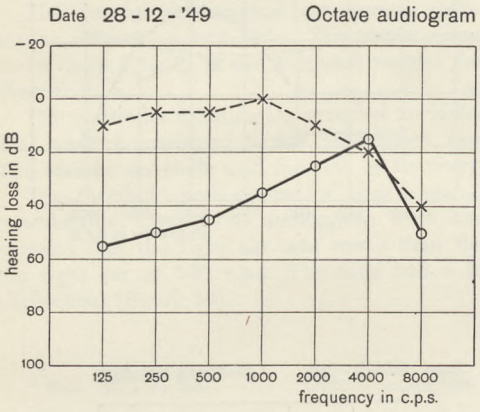
of the caloric test did not reveal a difference either in the irritability of both labyrinths. The marche en étoile and the tests of Romberg and Unterberger did not show any abnormalities.

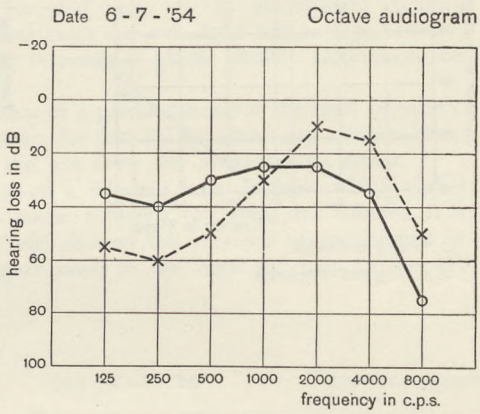
The threshold audiograms for pure tones showed a pronounced bass deafness in the right ear. This deafness had disappeared completely 3 days after the first examination but came back later on. Fowler's test (binaural balance) showed the presence of recruitment in the right ear at 500 c.p.s. The tone had a higher pitch in the left ear than in the right (diplacusis). A continuous audiogram was also made (figure 56).

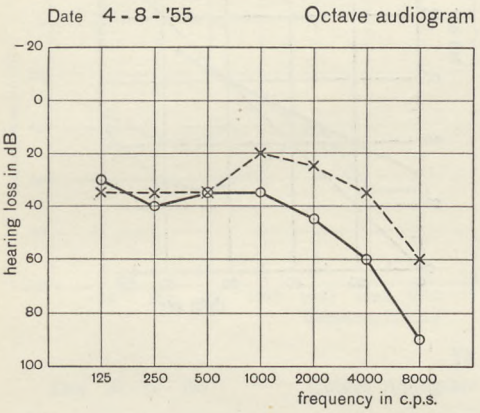
Case 15

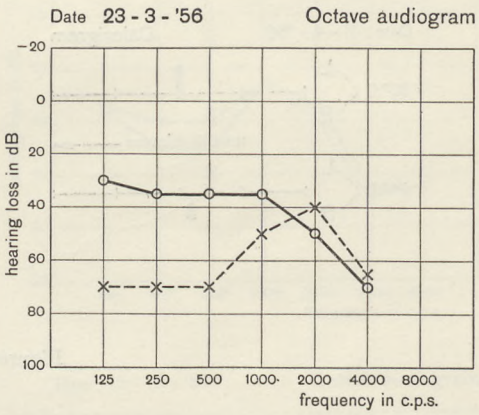
J. M. B.-P., woman, born 1888, housewife.

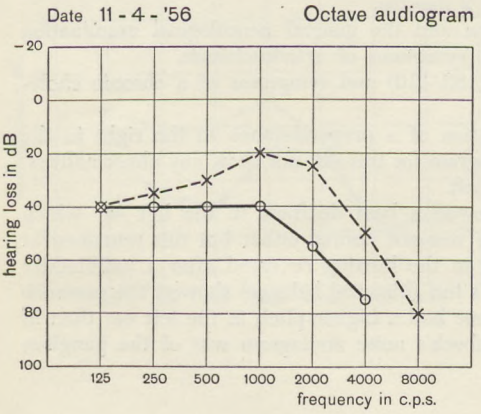
The patient was examined by us for the first time on 28-12-1949. She had complained of deafness and tinnitus in the right ear for 2 months. The audiogram that was made on 28-12-1949 showed a bass deafness in this ear.

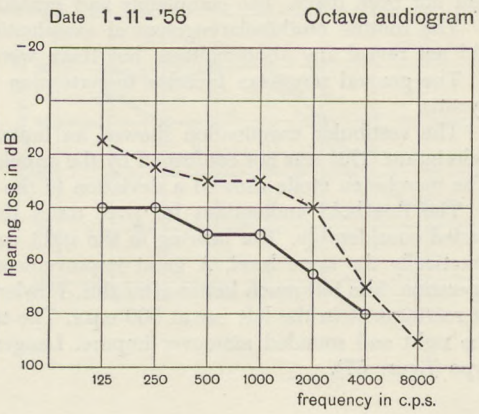












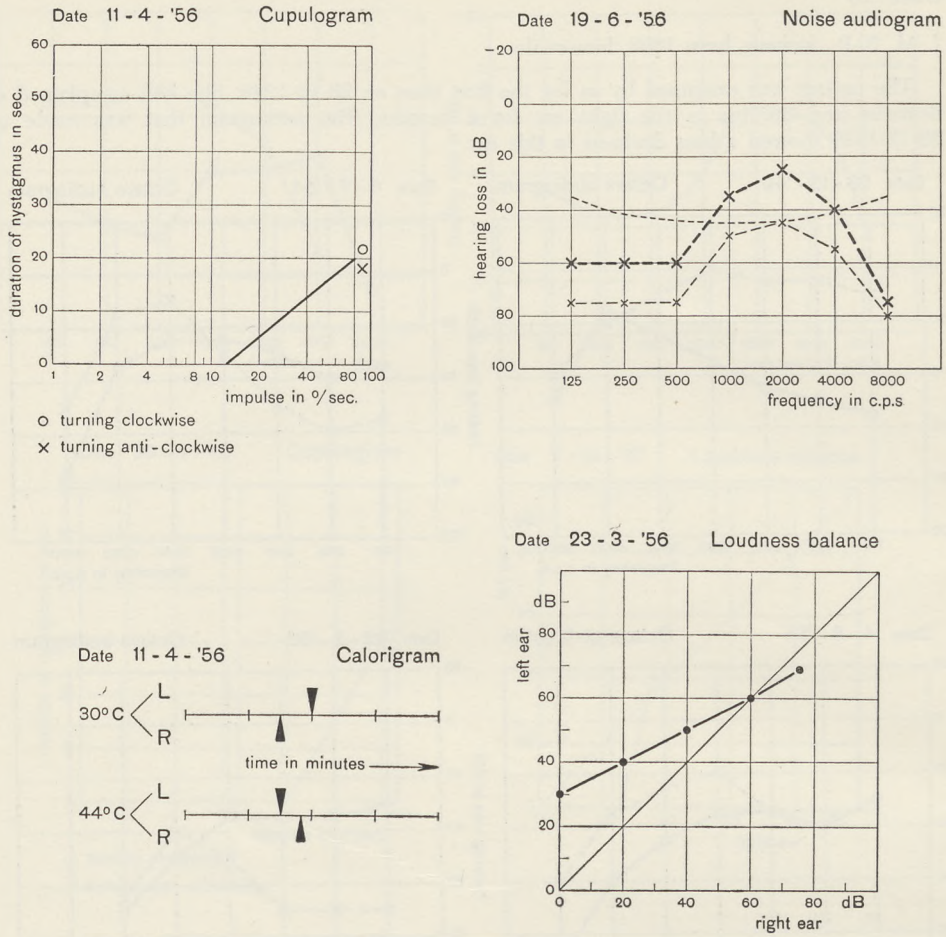


Figure 57

On 6-7-1954 she returned with complaints of deafness and tinnitus in the left ear. She had not been dizzy. The complaints had existed 2 months.

The routine otorhinolaryngological examination and the general neurological examination did not reveal any abnormalities, but there were symptoms of arteriosclerosis.

The general physician found a hypertension (180/110) and symptoms of a chronic cholecystitis.

The vestibular examination showed an indication of a preponderance to the right in the calorigram. This was not confirmed by the cupulogram for this did not show any abnormalities. The marche en étoile showed a deviation to the left.

The threshold audiograms for pure tones showed a bass deafness in the left ear which varied considerably. The hearing in the right ear was not normal either but this remained at practically the same level. A great improvement in the hearing occurred after a gallbladder operation. She felt much better after this. Fowler's test (binaural balance) showed the presence of recruitment in the left ear at 500 c.p.s. The tone had a higher pitch in the left ear than in the right and sounded moreover impure. Langenbeck's noise audiogram was of the ganglion type (figure 57).

Case 16

M. V.-van W., woman, born 1899, housewife.

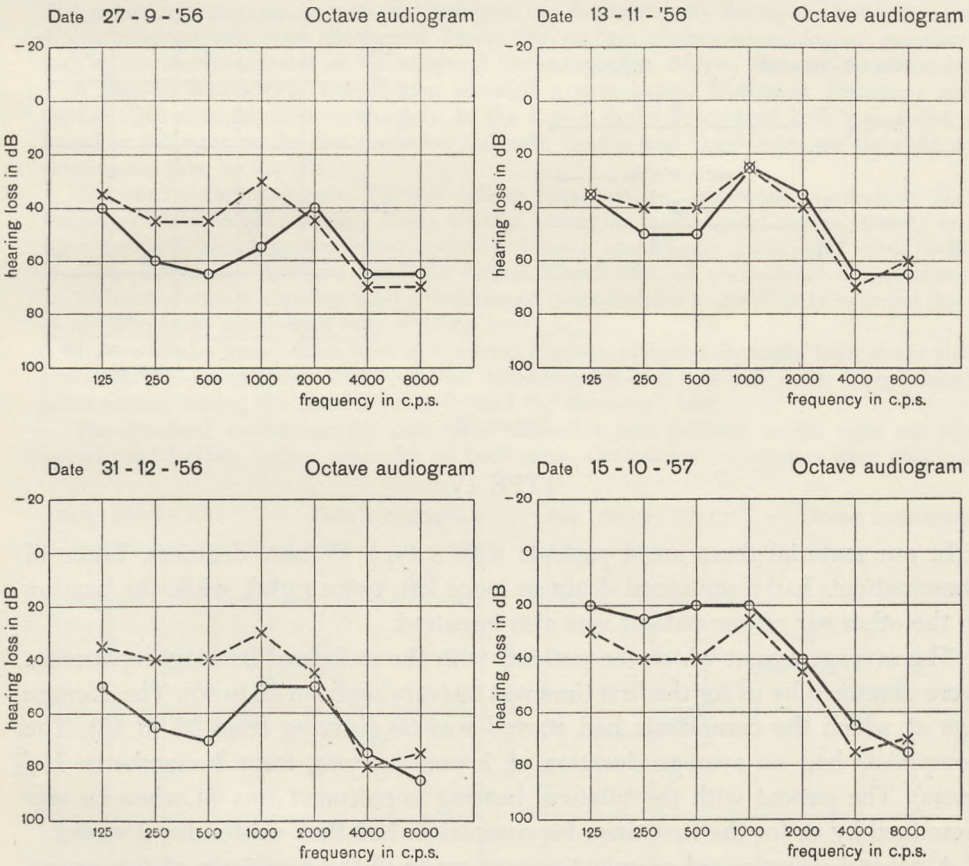
The patient was for the first time examined by us on 26-9-1956. She had complained of deafness and tinnitus in the right ear for 6 weeks. The hearing on both sides had been impaired for 1 year. She did not complain of vertigo.

The routine otorhinolaryngological examination did not show any abnormalities.

The general neurological examination only revealed very low to non-existent Achilles tendon reflexes on both sides. The electro-encephalogram showed slight diffuse beta activity. Some theta activity in the temporal region. Some response to photic stimuli. Conclusion: no abnormalities.

The vestibular examination revealed an indication of a predominance of the right labyrinth. This was not confirmed by the cupulogram, however, for this did not show any abnormalities. The marche en étoile and the test of Romberg did not show any abnormalities either.

The threshold audiograms for pure tones showed a bilateral bass deafness which varied considerably. Continuous audiograms were also made. Fowler's test (binaural balance at a moment that the right ear was worse than the left) showed the presence of recruitment in the right ear at 500 c.p.s. The tone had a higher pitch in the right ear than in the left (diplacusis) (figure 58).



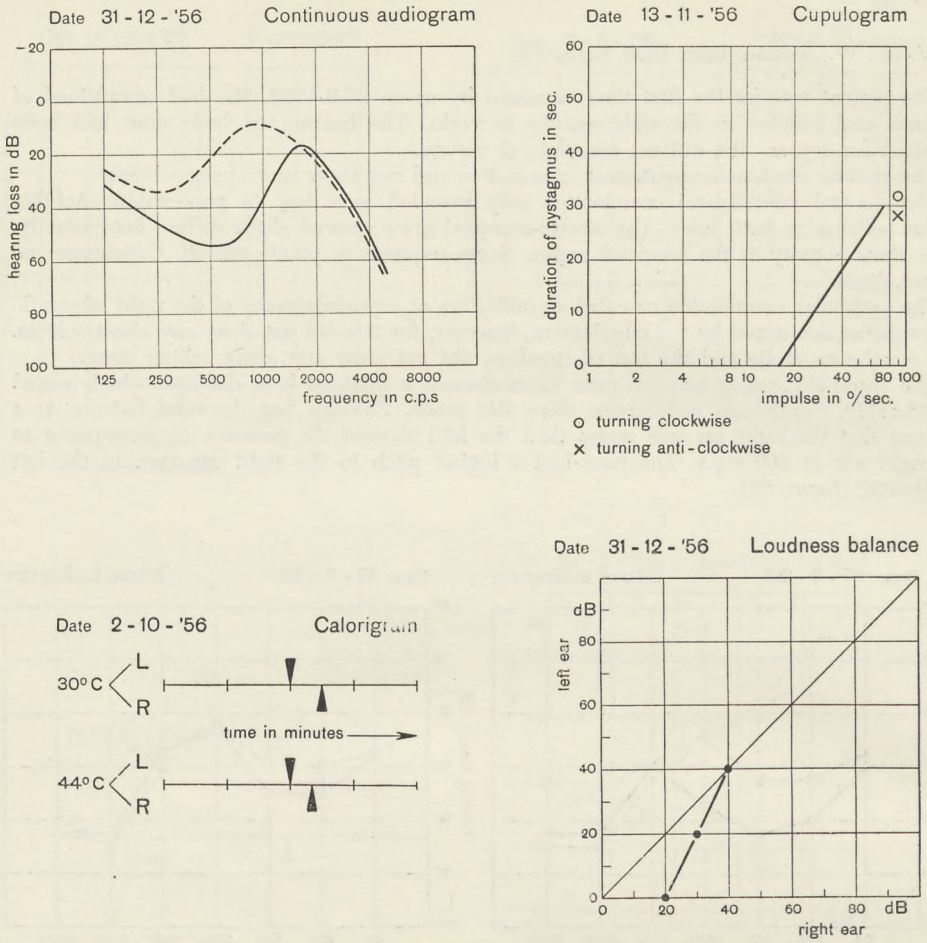


Figure 58

TYPE IV

In our material there are 4 patients with a type IV bass deafness. Three of these patients had a unilateral deafness (once left, twice right), while the hearing in the other ear of one patient was also impaired.

The average age at which the patients with the unilateral hearing impairment were examined by us for the first time was 39 (varying from 32 to 45). The average age at which the complaints had started was 38 (varying from 32 to 43). The complaints had an average duration of 1 year (varying from 4 months to 1½ years). The patient with the bilateral hearing impairment was 44 when he was examined by us for the first time; his complaints had then existed for 2 weeks.

A general neurological examination was made of the 4 patients of this group. Abnormalities were found in 3 cases. No diagnosis could be arrived at in any of these cases.

In 4 cases an electro-encephalogram was made, 2 of which showed abnormalities.

The 3 patients with the unilateral deafness were subjected to a vestibular examination. One of these, who had no complaints of vertigo, showed no abnormalities. Another patient who had no complaints of disturbances in his equilibrium either, showed a preponderance to the nonaffected side. The third patient who did have complaints of disturbances in his equilibrium, also showed a preponderance to the nonaffected side. The patient who also had hearing impairment on the other side, showed a predominance of the labyrinth on the side of the bass deafness.

Case 17

J. M. M., man, born 1917, instrumentmaker.

The patient was for the first time examined by us on 27-2-1957. He had had complaints of deafness and tinnitus in the right ear for 8 months. There were no complaints of vertigo. The patient was working in noise and had used ear defenders only during the last few months.

No abnormalities were discovered during the routine otorhinolaryngological examination and in the roentgenograms of the temporal bones (Schüller, Mayer, Stenvers and Steenhuis).

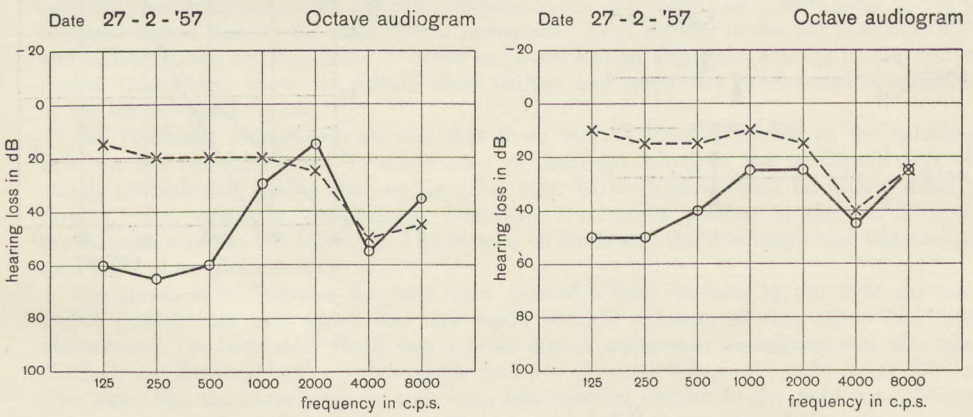
A general neurological examination revealed a pathological Hoffmann Trömner's reflex, on the right stronger than on the left. In the legs a slightly increased knee jerk reflex was found on the right; in the feet a positive Babinski, Gordon and Oppenheim, on the right more pronounced than on the left.

The electro-encephalogram showed diffuse beta activity, somewhat dominating in the temporo-parieto-occipital region. Theta activity mainly in the temporal region. Mainly in the right temporo-frontal region hyperventilation caused sporadically paroxysmal theta activity of a low frequency. Response to photic stimuli. Conclusion: no pronounced abnormalities.

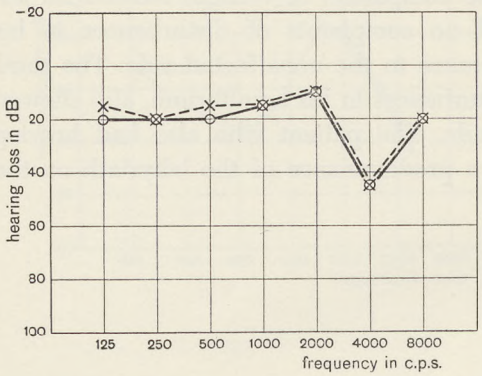
An internal examination revealed a pronounced exophthalmus and a slightly enlarged thyroid gland. The basal metabolism was + 11 %.

The vestibular examination showed a normal function of both labyrinths, both in the calorigram and the cupulogram. There was no difference between left and right. There were no abnormalities during the marche en étoile and the Romberg's test.

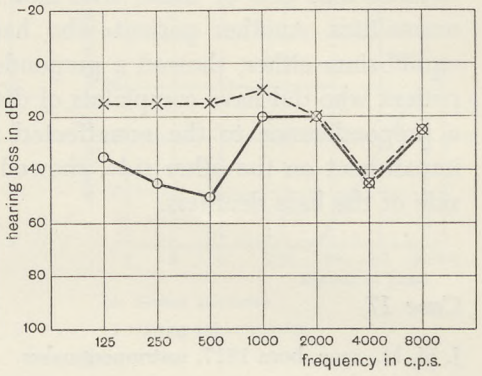
The threshold audiograms for pure tones showed a bass deafness in the right ear which varied considerably, with a noise dip on both sides. Continuous audiograms were also made.



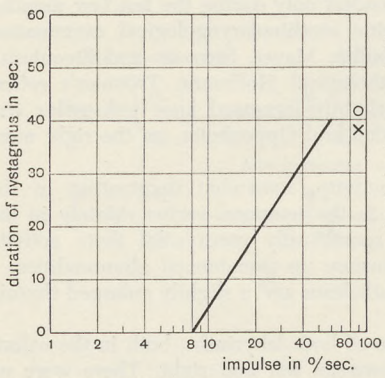
Date 11 - 3 - '57 Octave audiogram



Date 27 - 5 - '57 Octave audiogram

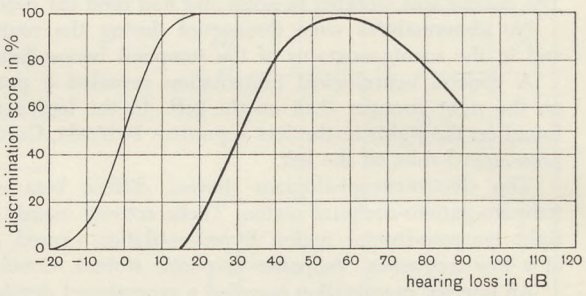


Date 11 - 3 - '57 Cupulogram

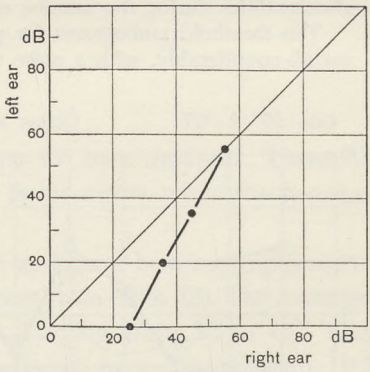


O turning clockwise
× turning anti-clockwise

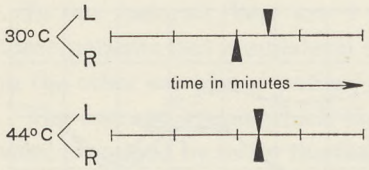
Date 7 - 1 - '58 Speech audiogram



Date 27 - 2 - '57 Loudness balance



Date 27 - 2 - '57 Calorigram



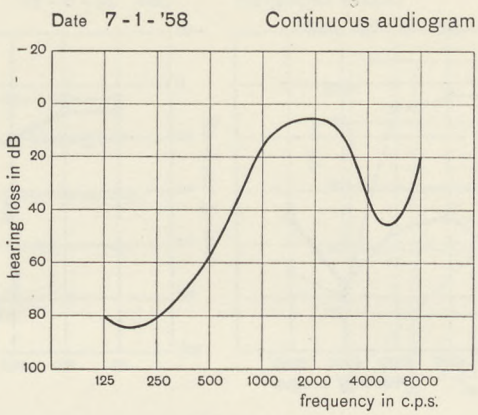


Figure 59

Fowler's test (binaural balance) showed the presence of recruitment in the right ear at 250 c.p.s. The tone had a higher pitch in the right than in the left ear (diplacusis). The speech audiogram showed a hearing loss of 30 dB in the right ear, while speech perception was 100 % when sufficiently amplified (figure 59).

Case 18

J. V., man, born 1925, draughtsman.

The patient was seen by us for the first time on 27-5-1957. He had suffered from deafness and tinnitus in the right ear for 4 months. From time to time he felt dizzy and sometimes he had to vomit during such a spell of vertigo. The patient was working in noise.

No abnormalities were found during the routine otorhinolaryngological examination. The roentgenograms of skull and temporal bones did not reveal any abnormalities either.

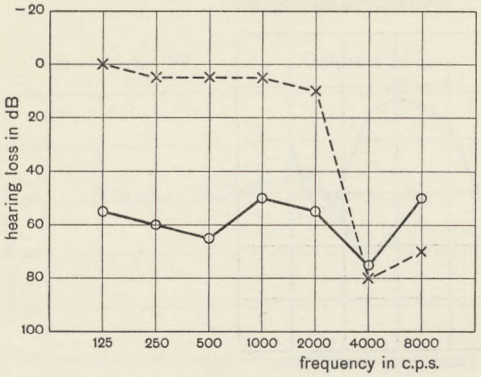
The general neurological examination revealed a very lively symmetrical picture of reflexes. In the right leg there was an expansion of the reflexogenic zone. On both sides there was a weakly positive Hoffmann Trömner's reflex.

The electro-encephalogram showed rather little alpha rhythm. In the occipital region, a little more on the left than on the right, beta activity of a low frequency. Some diffuse theta activity, predominantly in the temporo-occipital region and sometimes a little more in the left temporal region than in the right. Slight paroxysmal sharp activity in the left temporal region and isolated spike activity. Some response to photic stimuli and spike activity in the occipital region. Conclusion: there are diffuse abnormalities and slight but pronounced abnormalities in the left temporal region.

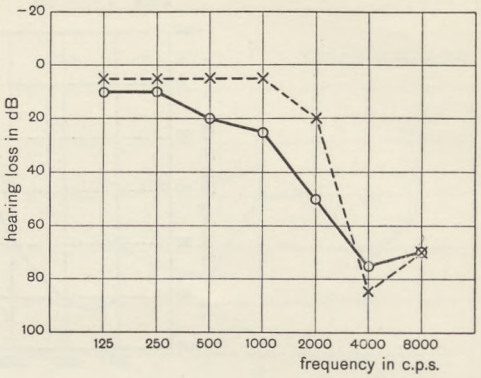
The vestibular examination showed that there was no clear difference in the calorigram between the two labyrinths; on calorization of the right labyrinth the nystagmus was of a clearly less splendid quality than on the other side. No nystagmus could be caused when the turning chair turned in anticlockwise direction. The results pointed in the direction of a predominance of the left labyrinth. The marche en étoile and the Romberg's and Unterberger's tests showed no abnormalities.

The threshold audiograms for pure tones showed a bass deafness in the right ear which varied considerably and which was now again strongly pronounced then again had totally disappeared. On both sides there was a noise dip. A continuous audiogram was also made. Fowler's test (binaural balance) showed the presence of recruitment in the right ear at 500 c.p.s. The tones had the same pitch in both ears, but sounded impure in the right ear. Monaural balance in the left ear from 2000 to 4000 c.p.s. revealed that recruitment was also present at the latter frequency (figure 60).

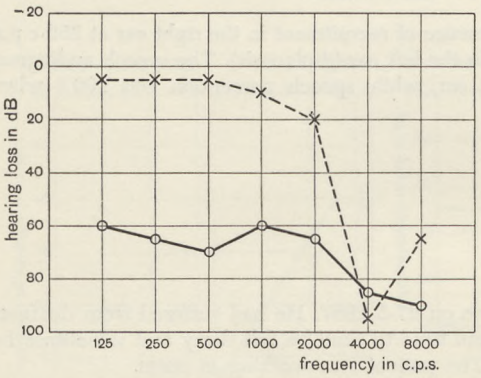
Date 28 - 5 - '57 Octave audiogram



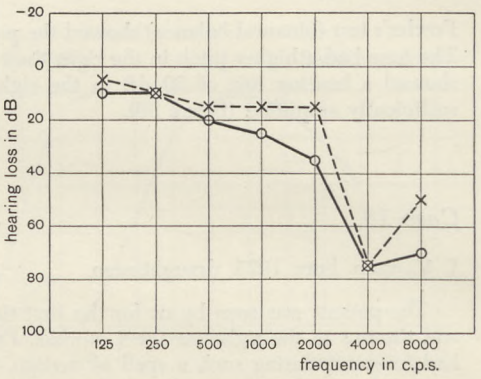
Date 25 - 6 - '57 Octave audiogram



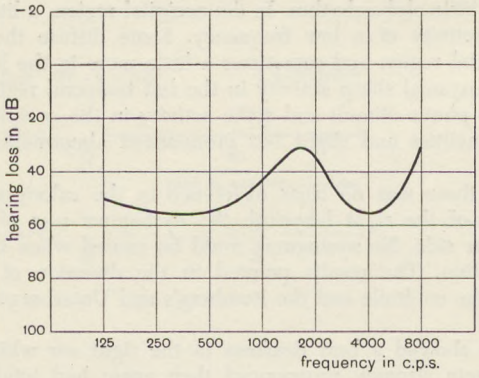
Date 16 - 7 - '57 Octave audiogram



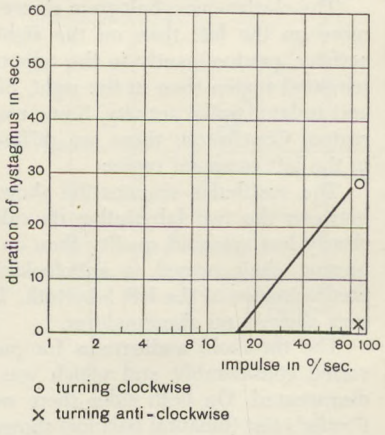
Date 24 - 9 - '57 Octave audiogram

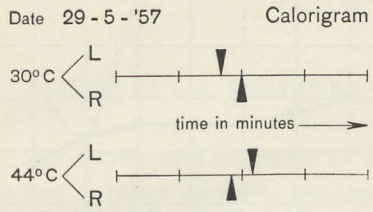


Date 29 - 5 - '57 Continuous audiogram



Date 29 - 5 - '57 Cupulogram





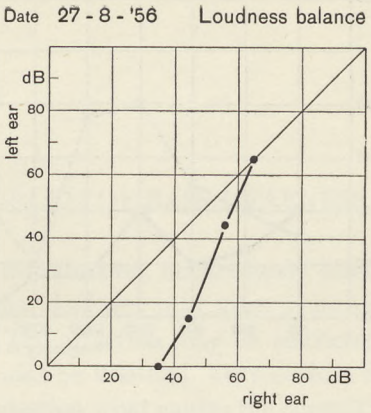


Figure 60

Case 19

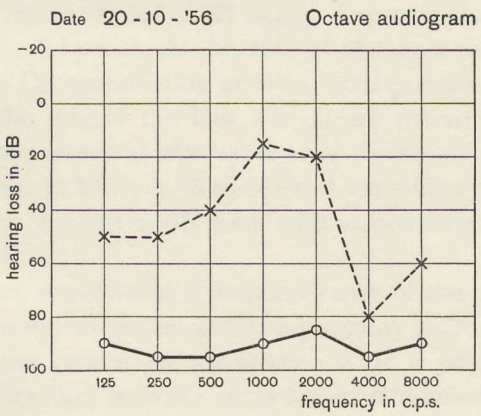
B. H., man, born 1912, factoryhand.

The patient was for the first time examined by us on 20-10-1956. He had complained of deafness and tinnitus in the left ear for a few weeks. Sometimes he was also dizzy for a moment. The patient worked in noise.

A routine otorhinolaryngological examination and the roentgenograms of the skull and the temporal bones showed no abnormalities.

The general neurological examination revealed a degenerative habit. The roentgenogram of the cervical spinal column showed an anomaly round the foramen magnum, which might lead to disturbances in the region of the vertebral artery. In the right arm and the right leg a varying rigidity with cogwheel phenomenon was found.

The electro-encephalogram showed diffuse paroxysmal spike and wave activity, predominantly in the temporo-frontal region, now a little greater on the left, then again on the right. Slight spike activity predominantly in the frontal region, sometimes more on the left than on the right. Some harmonic response to photic stimuli. Conclusion: there are diffuse epileptic abnormalities.



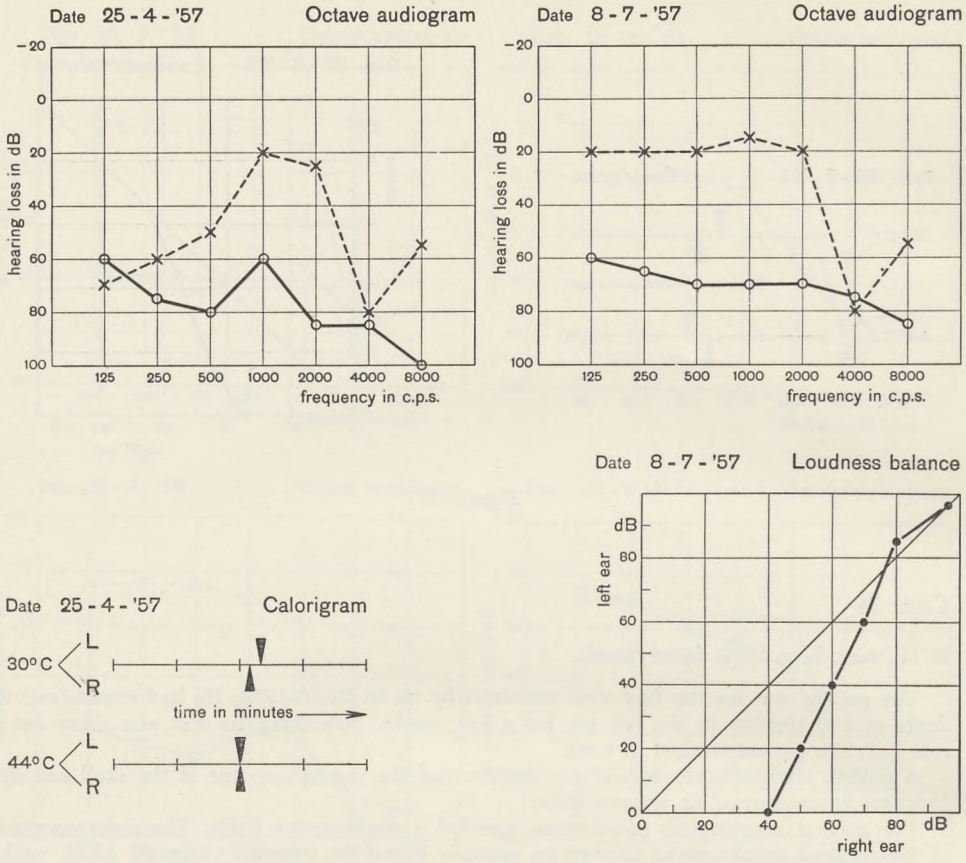


Figure 61

The first caloric test showed no difference in irritability of both labyrinths. A repetition of this test showed that there was a pronounced difference to the detriment of the right: predominance of the left labyrinth.

The threshold audiograms for pure tones showed a bass deafness in the left ear which varied considerably; the right ear had a flat hearing loss, fluctuating round 80 dB. Fowler's test (binaural balance) showed the presence of recruitment in the right ear at 2000 c.p.s. (figure 61).

Chapter IX

SOME FINAL REMARKS ON THE PROBLEM OF BASS DEAFNESS

Perceptive bass deafness, as defined in the introduction, occurs fairly often. It is just as often found in women as in men. Bass deafness may arise at any age, but it is especially found between 20 and 60. The affection may be restricted to the left or the right ear, but the deafness may also be bilateral. Very seldom does the anamnesis present a starting point for the question what causes the complaints. Nor do the internal, ophthalmic, roentgenological and neurological examinations reveal any specific abnormalities which may be directly connected with bass deafness.

As to the organ of equilibrium, which is so closely connected with the auditory organ, the findings vary considerably in cases of unilateral bass deafness. Often there are no complaints at all of disturbances in the equilibrium and quite often there are no abnormalities in the labyrinthine function. If abnormalities are found, they may be of the nature of a predominance of the labyrinth on the side of the nonaffected ear. Nearly as often a preponderance of the nystagmus to the normal side is found, while it sometimes occurs that there is a preponderance of the nystagmus to the affected side.

These findings are of a similar nature in case of bilateral hearing impairment. If there are abnormalities of the labyrinthine function they are mostly of the nature of a predominance of the labyrinth on the side of the bass deafness or of a preponderance of the nystagmus to the side of the bass deafness. The ear with the bass deafness is mostly the best ear in these cases.

Roughly speaking we may say that in approximately half the number of patients suffering from bass deafness, no abnormalities in the labyrinthine function are found (in our material in 20 out of 45 cases). Approximately a third part has a predominance of the labyrinth on the side of the best ear (in our material in 15 out of 45 cases). On examination about a sixth part has a preponderance of the nystagmus to the side of the best ear (in our material 8 out of 45 cases). Sometimes a preponderance of the nystagmus to the side of the worst ear is found (in our material in 1 out of 45 cases) and sometimes a spontaneous nystagmus (in our material in 1 out of 45 cases with a spontaneous nyastagmus to the side of the best ear).

Many investigators assume that a preponderance of the nystagmus points to a central cause of the abnormalities in the equilibrial functions, so a cause lying in the central nervous system. In our opinion there is no reason whatsoever to dismiss the possibility that in many cases too a predominance of one labyrinth might be based on a central disturbance.

We do not intend to go further into the problems that are put before us when trying to explain the results of the vestibular examination of our patients. This is a problem in itself which otherwise shows curious resemblances with the problem of bass deafness.

In the electro-encephalograms of patients with bass deafness we often see abnormalities, mostly of the nature of diffuse or isolated paroxysmal activity of an epileptiformic character. In our material such abnormalities were found in 16 out of 23 electro-encephalograms. The meaning of this is uncertain. Much more material will have to be collected on this point to be able to establish whether the electro-encephalogram can give information about the nature of a certain form of deafness. The abnormalities found would fit within the framework of the hypothesis established in chapter VI, regarding the hyperactivity of the efferent auditory-pathway system.

The tests to determine the accuracy of directional hearing in general do not give any results which give us a deeper insight into the nature of the disturbance on which the bass deafness found is based. In our material a central disturbance was only made likely in one case.

Confining ourselves to the findings in the auditory organ, we may ask ourselves the question what the picture of bass deafness looks like. In the first place there are the four types which have been described in the introduction. These types, as far as can be established, do not distinguish themselves in any fundamental way from each other.

The deafness is often peracute, without any occasion, and may, at least in the beginning, vary considerably. It occurs that a patient complaining of deafness comes during surgery hours and that an audiogram which is made during these very surgery hours, shows a normal hearing curve. Some days later the deafness returns and can also be seen in the audiogram. The hearing may vary considerably; now we find a hearing loss of 65 dB, then again a normal hearing. Smaller variations also occur.

Mostly only one ear is affected, sometimes the two ears alternate. It is just as if the hearing in the bass region „breathes". There is no regularity to be found in these variations and they grow less as the affection has existed longer.

Bass deafness is mostly attended with tinnitus, sometimes so violent that it is almost unbearable. This tinnitus is often of a low, buzzing character.

As has already been observed in chapter VIII a tone is appreciated differently by the affected and the unaffected ear. In the affected ear it sounds higher or lower, distorted or impure.

The recruitment phenomenon is constantly present.

Langenbeck's noise audiogram is of the ganglion type.

In the speech audiogram a surprisingly high percentage of the test words is often understood correctly when amplified optimally. The curve in the speech audiogram is often shifted far less to the right than might be expected on the strength of the tone audiogram. The curve mostly shows an optimum and then curls over downwards, as will happen in cases with recruitment.

In the continuous audiogram we find a curve which shows a remarkably steep slope on the side of the high frequencies.

On the face of these various findings it seems as if they are contradictory. The recruitment symptom, which finds expression in the balance test and the speech audiogram, is considered by many to be pathognomonic for an affection in the cochlea. The same holds good for diplacusis. Langenbeck's noise audiogram, however, is of the ganglion type. We have also made it plausible in the preceding chapters that the bass deafness described by us is never based on cochlear pathology in man.

In order to explain this seeming contradiction, it is necessary to review the threshold audiogram found. We find a curve which mostly indicates the greatest hearing loss for the frequencies 250 and 500 c.p.s. The hearing loss for 125 c.p.s. is mostly a little less great, while this loss to the side of the high frequencies mostly decreases quickly. This decrease strikingly finds expression in the type I continuous audiograms, in which, going upwards in the frequency scale, the hearing loss often decreases in the course of less than an octave from e.g. 60 dB to a negligible magnitude.

We should ask ourselves the question whether the threshold audiogram does give a real picture of the hearing loss present. For, both in the audiometer apparatus and the auditory organ itself harmonics may arise owing to alinear distortion. It is possible that the patient may hear these harmonics without being able to hear the fundamental.

In this connection FLETCHER's measurements are important. According to the method of the best beats, FLETCHER determined the intensity of the harmonics formed in the ear, in dependence on the intensity of the pure fundamental offered to the ear. In figure 62 this relation is rendered, valid for all frequencies.

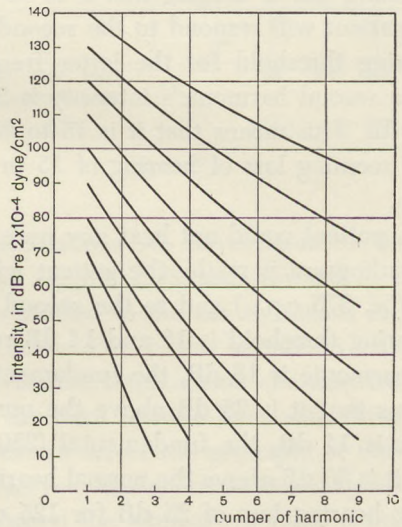


Figure 62

The intensities of whole series of harmonics, every time belonging to a certain intensity of the fundamental, may be read off. The intensity is indicated in dB re 2×10^{-4} dyne/cm^2, so in an absolute measure.

In order to establish a connection between these values and the hearing threshold, we must avail ourselves of the hearing threshold curve, also plotted out in an absolute measure. The simplest way is to take SIVIAN AND WHITE's minimum audible pressure curve for this. This is rendered in figure 63. We are aware of the fact that this threshold curve does not correspond entirely with the hearing threshold as found with an audiometer, because no allowance has been made for the influence of the auricle and the auditory meatus. This detracts nothing from the principle of the arguments, however.

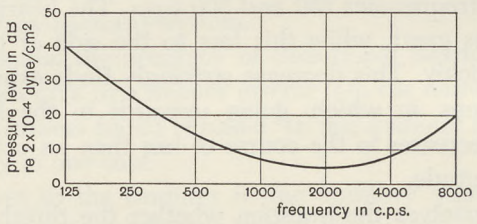


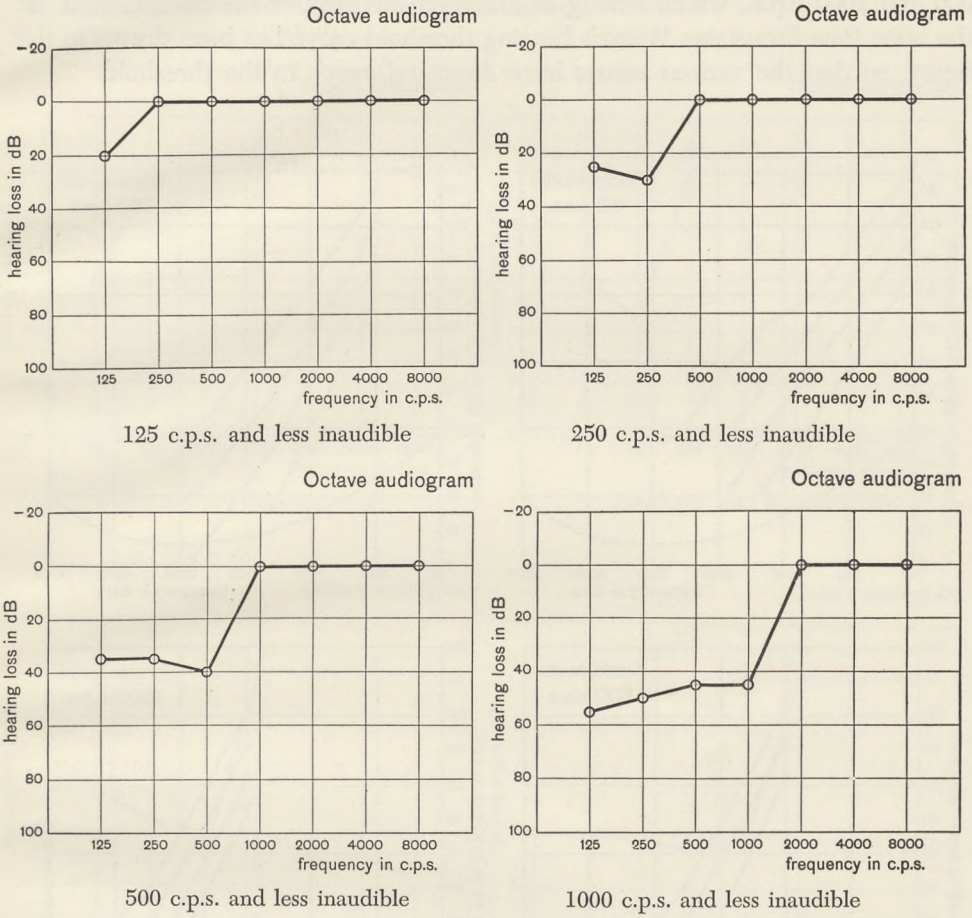
Figure 63

With the data collected in the two above figures we are entitled to assume what follows.

Let us suppose that a patient could not hear any more pure tones of 125 c.p.s. and less. When an octave audiogram would be made, he would still respond to a tone of 125 c.p.s. when this tone was so loud that the second harmonic would be audible for him. In the audiogram a hearing loss of a slight degree would be registered for 125 c.p.s., whereas the ear would in reality be completely deaf for this frequency. The seeming loss of hearing that is to be expected can be determined as follows. The patient will respond to the second harmonic of 125 c.p.s. i.e. 250 c.p.s. The hearing threshold for the latter frequency is 25 dB in an absolute measure. If the second harmonic's intensity is 25 dB, the fundamental itself must be 55 or 60 dB. This means that it is 15 to 20 dB above the normal hearing threshold. So a seeming loss of hearing of 15 to 20 dB for 125 c.p.s. is found in the audiogram.

Let us suppose that a patient could not hear any pure tones of 250 c.p.s. and less. When an octave audiogram is made, the patient will respond to the third harmonic of 125 c.p.s. (i.e. 375 c.p.s.) and to the second harmonic of 250 c.p.s. (i.e. 500 c.p.s.). The hearing threshold is 18 and 14 dB respectively for 375 and 500 c.p.s. If the third harmonic is 18 dB, the fundamental (125 c.p.s.) must be about 68 dB. This means that it is 25 dB above the normal hearing threshold. If the second harmonic is 14 dB, the fundamental (250 c.p.s.) must be about 55 dB. This means that it is 30 dB above the normal hearing threshold. So in this case we find a seeming hearing loss of 25 dB for 125 c.p.s. and of 30 dB for 250 c.p.s.

In this way the apparent hearing losses may be determined when it is supposed that the patient cannot hear pure tones of 500 and 1000 c.p.s. and less any more. In figure 64 the seeming hearing losses are rendered which may be expected in octave audiometry.



125 c.p.s. and less inaudible

250 c.p.s. and less inaudible

500 c.p.s. and less inaudible

1000 c.p.s. and less inaudible

Figure 64

While making these calculations we have assumed that the same hearing threshold curve holds good for the harmonics formed in the auditory organ as well as for the pure tones offered to the ear by means of the audiometer. This need not be the case of course. For, if the harmonics arise in the internal ear, they have nothing to do with the transmission characteristics of the middle ear. The tones offered to the ear from outside have something to do with them of course. This, however, does not affect the principle of the argument either.

As has already been observed above, harmonics may also arise in the audiometer apparatus. We measured the intensity of these distortion products arising in Peekel's audiometer. For this purpose we availed ourselves of an artificial

ear and a 1/3 octave band filter of Bruel and Kjaer, behind which was a frequency analyzer of General Radio with a band width of 6 c.p.s. Soon it proved that the odd harmonics were louder than the even harmonics. In the undermentioned figure only the odd harmonics have been plotted (figure 65). In this figure we can see the intensity of the harmonics for the frequencies 125, 250, 500 and 1000 c.p.s., which belong to a certain intensity of the fundamental. At the same time SIVIAN AND WHITE'S hearing threshold curve has been drawn in the figure, so that the various values have direct reference to the threshold.

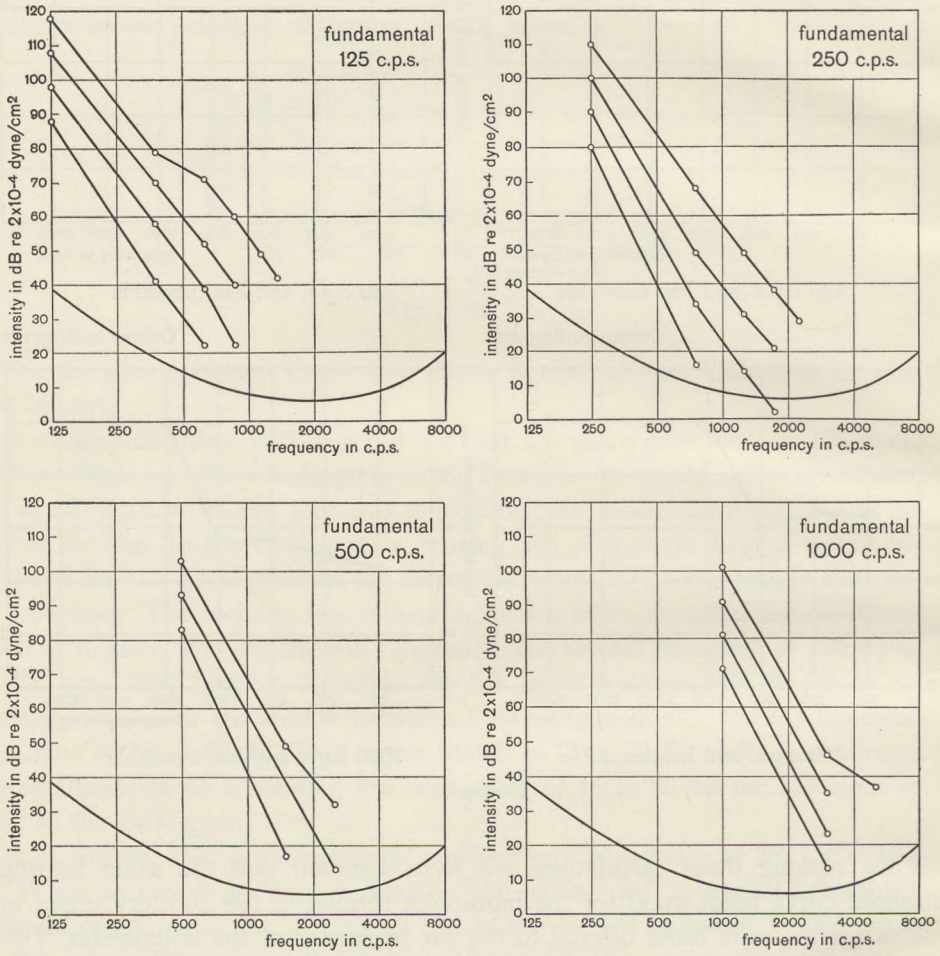


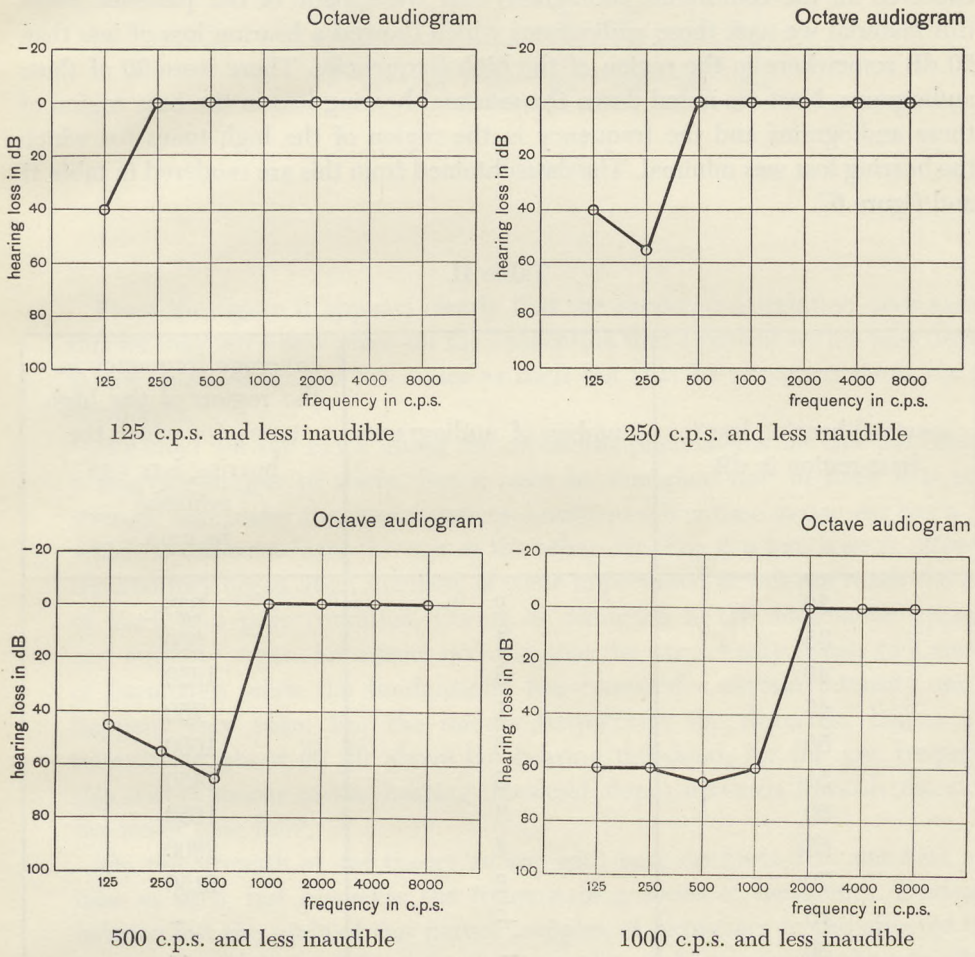
Figure 65

Now it is again possible to make similar suppositions as above.

Let us suppose that a patient cannot hear pure tones of 500 c.p.s .and less any more. He will respond to the fifth harmonic of 125 c.p.s., to the third harmonic of 250 c.p.s. and to the third harmonic of 500 c.p.s., when the octave audiogram

is made. If these harmonics should be just audible for the patient, the fundamentals should be about 83, 80 and 78 dB or in other words 45, 55 and 65 dB above the hearing threshold. These are the apparent hearing losses which are found in the octave audiogram for these frequencies.

These apparent hearing losses may also be calculated for other suppositions. In figure 66 we have drawn some possibilities.



125 c.p.s. and less inaudible

250 c.p.s. and less inaudible

500 c.p.s. and less inaudible

1000 c.p.s. and less inaudible

Figure 66

It is as yet difficult to decide in our patients whether the audiograms found show the greatest resemblance with the seeming hearing losses as a result of distortion in the auditory organ or with the seeming hearing losses as a result of distortion in the audiometer apparatus. Sometimes the first possibility seems most likely and sometimes the second.

However this may be, the resemblances between the hearing losses found and

calculated are striking enough to assume with a great measure of probability that the audiograms made of our patients do not give a real picture of the hearing loss present. We have reasons to suppose that a patient with bass deafness of the type described by us is in reality deaf for the low tones.

It is obvious that, on the strength of the above speculations, a correlation will be expected between the magnitude of the hearing loss in the bass region and the lowest frequency that can be heard normally again. In order to trace this we collected all the continuous audiograms that were made of our patients. From this material we took those audiograms which showed a hearing loss of less than 20 dB somewhere in the region of the high frequencies. There were 30 of these audiograms. Next we noted down the maximal hearing loss in the bass region of these audiograms and the frequency in the region of the high tones for which the hearing loss was minimal. The data obtained from this are rendered in table II and figure 67.

Table II

maximal hearing loss in bass region in dB	number of audiograms	average frequency in the region of the high tones, for which the hearing loss was minimal in c.p.s.
30	2	625
35	1	800
40	3	1250
45	1	750
50	6	1600
55	5	1450
60	3	1650
65	4	2800
70	2	2150
75	—	—
80	—	—
85	3	5350

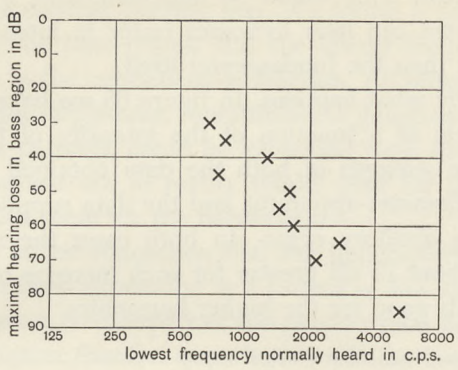


Figure 67

From the figure it appears clearly that the expected correlation does exist. In this we may see a new proof for the conception that a patient with a bass deafness is not able to hear the low tones as such but that he perceives them via their harmonics.

We shall not say much about the diplacusis phenomenon as this too could be a separate subject of study. But it must be remarked that in itself it is not so strange, that under the circumstances described a low tone in one ear has a pitch which is different from the one in the other ear. For if a low tone is offered to the auditory organ at an intensity of some importance, it will not reach the organ of Corti in a pure condition. Owing to distortion in the audiometer apparatus and auditory organ, harmonics occur so that the ear actually listens to a complex of harmonics when the fundamental has exceeded a certain intensity and this happens very soon. For the middle frequencies the distortion begins at an intensity of about 60 dB above the hearing threshold; for the low frequencies this occurs sooner as the hearing threshold slopes upwards towards the side of the lower tone limit.

On the strength of our theory an ear with bass deafness does not hear a low tone as such, but perceives the topmost of a series of harmonics. During the balance test the pitch of this partial complex of harmonics in the affected ear is compared with the pitch of the full complex of harmonics in the nonaffected ear. As the fundamental is loudest in the nonaffected ear, it will predominate there and determine the impression of pitch for the greater part. In the affected ear the fundamental is lacking and the incomplete complex of harmonics sometimes seems higher and then again lower in pitch than the complete complex. Nearly always the incomplete complex sounds highly impure and more so than the complete one, presumably also on account of the lacking of the predominating fundamental.

How can we explain that our patients show the recruitment phenomenon

during the balance test? This would be understandable if the complex of harmonics that the patient can hear increases faster in intensity when the fundamental grows louder than the fundamental itself.

This, now, is exactly what happens. In figure 68 we have plotted the intensity of the third harmonic as a function of the intensity of the fundamental. We have done so on the strength of both the data obtained when measuring the distortion of our audiometer apparatus and the data supplied by FLETCHER as to the distortion in the auditory organ. In both cases the intensity of the third harmonic becomes about 15 dB greater for each increase of the fundamental by 10 dB. This also holds good for the higher harmonics.

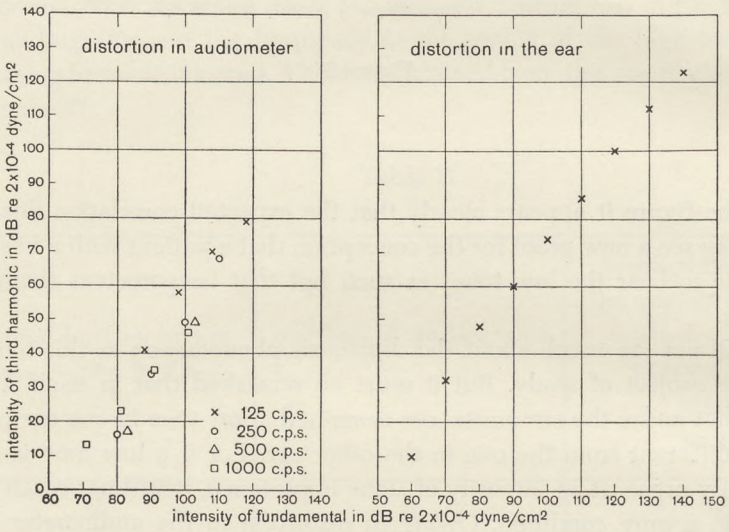


Figure 68

The results of Langenbeck's noise audiometry in patients with a bass deafness also become understandable now. For if such a patient can only perceive a low tone via the higher harmonics, this possibility of perception is lost for him when the higher harmonics are masked by a noise. We then see the remarkable phenomenon that a noise, which lies below the hearing threshold in the region of the low tones and consequently is not heard in this region at all, is yet able to raise the hearing threshold for low tones.

DISCUSSION

In chapter VI we already arrived at the conclusion that the inability to hear low tones might be explained from a condition of hyperactivity of the efferent auditory pathway system .Based on the data from the chapters VIII and IX it is possible to say something more about the way in which this hyperactivity finds expression.

We have seen that the threshold audiogram in cases of bass deafness does not

give a real picture of the existing hearing loss. We found a correlation between maximal hearing loss in the bass region and the lowest frequency which may be heard normally again in the continuous audiogram. All other symptoms in the field of the auditory organ (diplacusis, recruitment, a ganglion type of the noise audiogram) found in our patients, could be explained from the supposition that patients with a bass deafness are in reality totally deaf for the low tones and that they can only perceive them via the harmonics.

It is obvious to arrive at the conclusion that the hyperactivity in the efferent auditory pathway system finds expression in a raising of the lower tone limit.

So actually we have here a counterpart of the phenomenon described by GUILD (chapter IV). This investigator found a quantitative relation between the number of available nerve fibres in the cochlear nerve and the upper tone limit. Each decrease of the number of intact nerve fibres by 10 % lowers the upper tone limit by one octave. This is a purely anatomical relation; the hearing loss is always irreversible if the damage of the nerve fibres is irreparable.

The raising of the lower tone limit by hyperactivity of the efferent auditory pathway system must rather be considered as being functional. This fits in with the greatly varying character of bass deafness, the recovery of the hearing in the bass region sometimes even after several years and the phenomenon of the alternate occurrence of bass deafness in the right and the left ear.

Naturally we are not able to give further insight into the problem regarding the way in which the efferent auditory pathway system can influence the position of the lower tone limit. Much more work will have to be done, particularly in the field of neurophysiology, before we shall be able to say that the described mechanism will be known to us in detail.

About the therapy of bass deafness we have not spoken on purpose. We are not under the impression that the medicine prescribed by us (nicotinic acid, phenobarbital, etc.) have had any drastic influence on the course of the affection. It is also obvious that a rational therapy cannot be found before a better insight is gained into the processes which underlie the origin of bass deafness.

SUMMARY

Many investigators have thought for a long time that bass deafness could never be of a perceptive kind. If an audiogram revealed a hearing loss for the low tones that proved to be greater than that for the high tones, it was always said that this was due to an affection of the middle ear, or in other words the diagnosis in such a case was a conduction deafness. Many misunderstandings have arisen because insufficient precautions were taken while recording the bone conduction of the affected ear. This should always take place while the nonaffected ear is masked simultaneously.

If this requirement is conscientiously conformed to, we are struck by the fact that relatively bass deafness of a perceptive character occurs fairly often. The patient is then at the same time preserved from a prolonged treatment for tubal catarrh, an affection from which he is not suffering at all.

Perceptive bass deafness may find expression in the audiogram in various ways. Let us put first and foremost that we do not speak of bass deafness in cases in which the hearing loss for the lower tones is indeed greater than for the high tones, but the audiogram curve shows a very faint slope. For in that case, besides a hearing loss of some importance in the region of the low tones, there is also a hearing loss in the region of the high tones. It is plausible that in such cases the hearing loss may among other things be based on a disturbance in the hydrodynamics of the cochlea (endolymphatic hydrops, changes of mass and stiffness of the basilar membrane) or a disturbance in the supply of the hair cells from the surrounding labyrinthine fluid.

We prefer to reserve the term bass deafness for those cases in which the audiogram shows a hearing loss for the low tones, while the hearing in the middle region or in the region of the high tones is normal or approximately normal. This rule should be applied with some suppleness. For instance it is possible that a bass deafness arises in an auditory organ which is already impaired because of a hearing loss for the high tones e.g. owing to presbycusis or noise trauma. It may also occur that a hearing loss for the low and the high tones occurs at the same time. In both cases it is only obvious that there is among other things a bass deafness if the hearing in the middle region is relatively very good. Finally a bass deafness may be completely hidden by a hearing loss with a flat curve in the audiogram. Bass deafness cannot be diagnosed then and it only becomes plain that there was one after all, if the hearing in the bass region improves, independent of the hearing in the region of the high tones.

Continuous audiometry is an excellent method to establish whether there is a bass deafness or not, and if there is one, what form it has.

It is true, that it is possible to cause a bass deafness by damaging the apical

coils of the cochleas of test animals, but it is not likely that a bass deafness of the type described by us is ever based on cochlear pathology in man.

A lesion of the central auditory pathways may cause a bass deafness. It is practically impossible to explain the origin of bass deafness from an affection of the afferent auditory pathway system. It is possible, however, to find a natural explanation of the origin of bass deafness on the strength of the hypothesis that the efferent auditory pathway system, in a hyperactive condition, is able to give rise to a raising of the lower tone limit. In what way this raising is achieved, remains a problem as long as neurophysiology is unable to supply further information about this.

Patients with a bass deafness of the type described by us are actually totally deaf for the low tones; they react to the harmonics of these low tones while an audiogram is made. These harmonics arise because of alinear distortion in the audiometer apparatus or in the auditory organ. Consequently the audiograms of patients with bass deafness do not give a real picture of the existing hearing loss.

The audiological findings, according to current conceptions, now point in the direction of a cochlear focus, then again to a central focus of the affection on which the deafness is based. This discrepancy is only apparent and may be explained from the fact that the patient perceives harmonics instead of the fundamental and from the behaviour of these harmonics, which, when the intensity of the fundamental increases, increase in intensity and loudness at a much greater rate than this fundamental.

The clinical data of 45 patients, discussed in the preceding chapters, clearly show the picture of bass deafness. The syndrome shows a remarkable uniformity in spite of the fact that the deafness may vary considerably and the attendant phenomena, particularly the disturbances in the equilibrium, are not present in every case.

This syndrome is definitely not identical with Ménière's disease (endolymphatic hydrops), in spite of the fact that complaints of vertigo often occur in it.

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STELLINGEN

I

Kinderen die een normaal gehoor hebben voor de lage tonen, doch die niet of slecht in staat zijn de hoge tonen waar te nemen (kinderen met een zogenaamd diagonaal audiogram) behoren thuis op een school voor slechthorenden en spraakgebrekkigen.

II

De behandeling van de patiënt met asthma bronchiale door de heilgymnast toont grote overeenkomst met de logopaedische behandeling van de stotteraar. In beide gevallen dient de nadruk te worden gelegd op het ontspannende element in de voorgeschreven oefeningen.

III

De beantwoording van de vraag of de tonsillen een focus van infectie kunnen vormen, is voor de keel-, neus- en oorarts in concrete gevallen vaak onmogelijk en in het algemeen zeer moeilijk. Dit moet geweten worden aan het feit dat hij de tonsillectomie vaak uitvoert op verzoek van een ander specialisme en er geen gecoördineerd na-onderzoek plaatsvindt.

IV

Indien kunstmatige inseminatie bij de mens met sperma van een ander dan de echtgenoot moreel verantwoord en wettelijk geoorloofd zou worden geacht, zouden op genealogische en eugenetische gronden voldoende waarborgen dienen te worden geschapen ter beveiliging van de identificatie-mogelijkheid van de donor.

V

Het is onjuist te menen dat een slijmvliescyste in de sinus maxillaris aanleiding kan geven tot klachten over hoofdpijn. Operatieve verwijdering van een dergelijke cyste komt dan ook niet in aanmerking.

VI

De klinisch-audioloog zou ten zeerste gebaat zijn bij een nauwe samenwerking met andere specialisten, waaronder de patholoog-anatoom, opdat vaker dan thans het geval is, de bij patiënten tijdens het leven verkregen audiologische gegevens kunnen worden gecorreleerd met de post mortem gevonden afwijkingen in cochlea en gehoorbaan.

VII

Uit de Voorschriften voor het Ziekenfondswezen, punt 12 van het rondschrijven van 16 februari 1944, IVde Afd., Nr. 1767, betreffende de verstrekking van spraaklessen, dient de zinsnede „bijvoorbeeld na operatie van een gespleten verhemelte" te vervallen.

„Verplicht-verzekerden hebben in het algemeen geen recht op verstrekking van spraaklessen. Slechts indien deze lessen deel uitmaken van de medische behandeling, bijvoorbeeld na operatie van een gespleten verhemelte, kunnen spraaklessen voor rekening van het algemeen ziekenfonds worden gebracht. Verstrekking is afhankelijk van de machtiging van de controlerend geneesheer. Ingeval van twijfel, of behandeling voor fondsrekening komt, behoort de aangelegenheid aan mij (voorzitter ziekenfondsraad) ter beslissing te worden voorgelegd."

VIII

Bij de operatieve behandeling van een vliesvormige obstructie van het onderste gedeelte van de oesophagus („lower esophageal web" volgens Bugden) dient, zo mogelijk, de spierlaag gespaard te blijven en de ingreep beperkt te worden tot mucosa en submucosa.

IX

Indien in het kader van een strafrecht-procedure de geestestoestand van de verdachte moet worden beoordeeld door de psychiater, dient deze zich in zijn conclusie te onthouden van een uitspraak betreffende de mate van toerekeningsvatbaarheid. Hij beantwoorde de vraag naar de toerekeningsvatbaarheid onder deze omstandigheden slechts in positieve of negatieve zin.

X

Het bestaan van een abnormale correspondentie tussen de retinae van beide ogen kan bij de mens, in het bijzonder indien de afwijking gering is, het beste worden vastgesteld met behulp van het phenomeen van de Haidinger bundels.

1 juli 1958. D. W. GRAVENDEEL.