SUDDEN PERCEPTIVE DEAFNESS

TH. A. BIERMAN

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PROEFSCHRIFT

TER VERKRIJGING VAN DE GRAAD VAN DOCTOR IN DE GENEESKUNDE AAN DE RIJKSUNIVERSITEIT TE LEIDEN OP GEZAG VAN DE RECTOR MAGNIFICUS DR. S. E. DE JONGH, HOOGLERAAR IN DE FACULTEIT DER GENEESKUNDE, TEGEN DE BEDENKINGEN VAN DE FACULTEIT DER GENEESKUNDE TE VERDEDIGEN OP DONDERDAG 19 DECEMBER 1957 TE 14 UUR

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Aan mijn Ouders en Zusters
Aan mijn Vrouw en Kinderen

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CONTENTS

SAMENVATTING

Door de observatie in Februari 1954 van twee patiënten lijdende aan plotselinge perceptiedoofheid tesamen met meningeale prikkelingsverschijnselen en bij wie uit de faeces en liquor het Columbia S.K. virus werd geïsoleerd, kwamen wij op de gedachte dat virusinfectie in het algemeen bij deze vorm van doofheden weleens een belangrijk etiologisch moment zou kunnen zijn. Hierom werd systematisch naar virusinfectie gezocht.

Het is bekend dat intoxicaties en sommige infectieziekten als bof zulk een plotseling gehoorverlies als gevolg kunnen hebben. Eveneens komen plotselinge schommelingen in het gehoor bij de ziekte van Ménière regelmatig voor. Deze gevallen, waarbij dus de oorzaak bekend is, hebben wij in tegenstelling tot andere auteurs, zoals HALLBERG, niet in ons onderzoek betrokken.

Van de 110 patiënten, verzameld vanaf 1954 tot en met Augustus 1957, werden 74 door ons clinisch geobserveerd en 36 verkregen door middel van een enquête gehouden onder Nederlandse Keel-, Neus-, Oorartsen. De totale groep omvatte 68 mannen en 42 vrouwen, die een gemiddelde leeftijd hadden van 42 jaar.

Onder hen bevonden zich slechts 19 patiënten lijdende aan bilaterale perceptiedoofheid, waarvan 11 met een gehoorverlies aan beide oren van groter dan 80 dB, dat wil zeggen dat zij nagenoeg totaal doof waren. De resterende 8 patiënten waren in geringere mate doof voor spraak. Bij de 91 unilaterale doofheden was het verstaan van spraak niet gestoord omdat in het andere oor de gehoorscherpte steeds voldoende gebleven was. De graad van doofheid aan het betrokken oor varieerde van 30 dB. verlies tot totale uitval. Het regressie symptoom dat bij 51 patiënten werd nagegaan, was bij 29 van hen aanwezig. Wat de gehoorsfunctie betreft, werd een verbetering vastgesteld bij 45 patiënten, waaronder ook 2 met een bilaterale doofheid. Een neiging tot spontane verbetering kon bij 19 onbehandelde patiënten aangetoond worden.

Het zeer hinderlijke verschijnsel van tinnitus, waardoor soms de doofheid bemerkt wordt, was bij 78 patiënten aanwezig. De doofheid werd door dit symptoom voorafgegaan bij 17 van hen en werd slechts bij 5 pas later bemerkt. Bij ongeveer de helft van deze patiënten verdween de tinnitus binnen 1 maand en slechts in enkele van de overige gevallen was dit verschijnsel blijvend.

Evenals tinnitus, is diplacusis een symptoom van een cochleaire laesie. Dit verschijnsel werd door 73 patiënten, bij navraag, aangegeven. Bij 4 patiënten was diplacusis zelf de aanleiding tot het bemerken van hun doofheid. Ook dit symptoom is tijdelijk aanwezig en werd slechts door 2 patiënten als blijvend aangegeven.

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Duizeligheid kwam voor in de ziektegeschiedenis bij 52 van onze patiënten. Het calorisch onderzoek van het labyrinth werd verricht bij 71 patiënten, waaronder 10 bilaterale doofheden waren. Het resultaat van dit onderzoek was dat er bij 33 patiënten een functie-stoornis van het labyrinth werd aangetoond, dat varieerde tussen een lichte afwijking en totale uitval, terwijl er vaak een nystagmus-voorkeur aangetoond kon worden. Een spontane nystagmus werd slechts bij 9 patiënten lijdende aan unilaterale doofheid gezien. Merkwaardig was dat bij 3 patiënten, waarbij een subtotale doofheid aan beide oren werd gemeten, normaal reagerende labyrinthen werden aangetoond, terwijl bij 5 anderen slechts één labyrinth gestoord bleek te zijn.

In de gevallen, waar koorts met meningeale prikkelingsverschijnselen en een griepachtige infectie aanwezig was, werd ons vermoeden dat een virusinfectie de oorzaak zou kunnen zijn inderdaad bevestigd. Hoofdpijn en neuscatarrh waren bij hen vaak vroegsymptomen. Zo werden afwijkingen van de liquor cerebrospinalis, die wezen op een benigne sereuze meningitis, bij 14 van deze patiënten gevonden. Bij 74 patiënten waaronder 48 verdacht van een virusinfectie en bij wie het begin niet lang geleden was, werd virologisch onderzoek gedaan. Uit de faeces en de liquor werden bij 2 van hen virus gekweekt. In 25 gevallen waren de serologische reacties positief en wel in niet minder dan 14 gevallen voor het bofvirus. Slechts 1 patiënt had een lichte parotitis. Allen hadden reeds vroeger bof doorgemaakt zodat wij hier met reinfecties te maken hebben. Een contact met boflijders was slechts aan 6 patiënten bekend. Dit voorkomen van bofreacties in een derde van de gevallen van virusinfecties, is statistisch significant voor de oorzakelijke betekenis van het bofvirus. Immers door WOLFF is aangetoond dat onder normale proefpersonen positieve bofreacties slechts in $\pm 0{,}1\,\%$ voorkomen, terwijl onder mensen die in contact met boflijders geweest waren in 99,3 % positieve reacties gevonden worden.

Voor de 7 positieve reacties, aangetoond voor de „Enterale groep", namelijk 5 voor Columbia S.K. en 2 voor de Coxsackie-groep, is de causale samenhang minder zeker, echter toch waarschijnlijk.

Voor de influenza-groep werden 7 positieve reacties gevonden. Dit getal is zeker niet significant omdat onder normale proefpersonen eveneens een ongeveer gelijk aantal verwacht mag worden.

Door vroegere onderzoekers is de etiologie en pathogenese van de acute perceptiedoofheid vaak niet voldoende onderscheiden. Zo werd als oorzaak vaak aangenomen circulatiestoornissen en labyrinthaire hydrops, terwijl men toch moet aannemen dat aan deze pathogenetisch belangrijke mechanismen een diepere oorzaak ten grondslag ligt. Zo zullen bij infectie steeds oedeem en vaatafwijkingen gevonden worden. Slechts bij 12 van onze patiënten, die cardiovasculaire symptomen hadden, werd aan een vasculaire oorzaak zoals spasme en thrombose

gedacht. In ongeveer de helft van onze 110 patiënten bleef de etiologie onbekend. Zeer goed mogelijk is het, dat onder deze patiënten onbekende of niet herkende virusinfecties een belangrijk moment zijn.

Over de localisatie van het proces bij deze ziekte werd, waar regressie en diplacusis aanwezig was of perifere vestibulaire afwijkingen, een cochleaire laesie aangenomen. Dit sluit echter niet uit, dat, vooral in de gevallen van virusinfectie, een combinatie met een neuritis en encephalitis zeer waarschijnlijk is.

De therapie van de plotselinge perceptiedoofheid veroorzaakt door virusinfecties is machteloos. Aureomycine, salicylas natricus werden geprobeerd, echter zonder resultaat. Aangezien verondersteld werd dat, in vele gevallen en zelfs ook bij infectie, vaatspasmen een rol spelen werden spasmolytica, als nicotinezuur en aminophylline, toegediend. Audiometrisch gecontroleerde directe verbeteringen werden herhaaldelijk waargenomen. Deze verbeteringen waren echter meestal niet blijvend en indien blijvend moet met de mogelijkheid van spontane remissies rekening gehouden worden.

De prognose van deze plotselinge perceptiedoofheden is, voor wat de sociale validiteit betreft, niet zo slecht als men tot nu toe meende. Van onze 110 patiënten hadden 91 een unilaterale doofheid en van hen waren 53 absoluut doof. Van deze 53 herstelden 22, terwijl de overige 31 met hun normaal oor voldoende konden horen. Van de 19 patiënten met bilaterale doofheid herstelden 2 volledig en 6 zodanig dat zij met een hoorapparaat redelijk konden horen. Niet minder dan 11 bleven geheel doof. Dit is dus 10% van het totale aantal patiënten.

Naar schatting komt in Nederland per jaar een aantal van 200 van deze plotselinge perceptiedoofheden op een bevolking van elf millioen voor. Natuurlijk is het mogelijk dat bij epidemieën dit aantal tijdelijk zal toenemen.

INTRODUCTION

Deafness can be divided into two broad categories according to its origin. In the first of these categories the distinguishing feature is impairment of the conduction mechanism and in the second, of the perception mechanism. This group of perceptive deafness includes not only lesions of the organ of Corti but also affections of the VIIIth nerve, of the nucleic area and of the cerebral cortex. Another separate group is formed by the psychogenic forms of deafness. According to the underlying diseases, all these types can occur either gradually or very suddenly and they may also tend to grow worse in a short time.

This study deals with those types of perceptive deafness whose onset is sudden and whose cause is unknown. This automatically excludes perceptive deafness caused by trauma, intoxications (quinine, carbon monoxide, salicylates) and bacterial infection (meningitis, syphilis, etc.) Also excluded are the sudden exacerbations of deafness associated with an attack of Ménière's disease. Some authors (see Section II) include this type of deafness in their study on the ground that sudden attacks of deafness may all have the same pathogenesis: increase in pressure in the endolymphatic system. In addition, the sudden deafness of the patients with whom we are concerned is frequently accompanied by dizziness. However, the clinical picture presented by these two diseases shows such wide differences that a separation between them is certainly desirable. The results of this present investigation will indicate that this interpretation is justified.

We have included in this study only patients satisfying the following criteria:

1. It could be established with sufficient certainty that before the sudden loss the patient's hearing was good or moderate in the affected ear or ears.
2. This attack of deafness was the first experienced.
3. The hearing was so impaired by this attack that normal or loud conversation was no longer or barely understood.

Observation of a patient whose sudden attack of deafness was associated with symptoms of a "benign" lymphocytic meningitis led to the supposition that it was very possible that virus infections might play an unsuspectedly large role in this form of deafness. It has indeed long been known that a virus infection, such as mumps, can result in serious perceptive deafness. Our study was thus set up to discover, if possible, whether this group of patients could be shown to have suffered a virus infection. To this end we were able to secure the help of the Institute

for Tropical Medicine (Director, Prof. J. E. DINGER, M.D.) and of the Institute for Preventive Medicine (Director, Prof. J. D. VERLINDE, M.D.).

Starting with the supposition that deafness is only a symptom and that not only virus infections but other causes may underly the disease, in addition special investigations were carried out to determine whether such causes as vasospasm, thrombosis, and abnormal blood coagulation mechanism could be demonstrated.

I

REVIEW OF THE LITERATURE

There is very little literature concerning sudden perceptive deafness. Most of the existing publications discuss only a small number of patients, which makes the selection of the material very variable. As stated above, frequently cases of Ménière's disease and neurological disturbances are included in this category of deafness. Other publications include cases with a known cause.

So KOBRAK, 1922, described 7 cases caused by trauma and intoxication and only one case whose cause could not be given.

In 1934, COLLET presented a report on perceptive deafness to the Third Congress of the Societas Oto-Rhino-Laryngologica Latina. In this report it is assumed that deafness following mumps, typhus, grippe and especially herpes zoster oticum is caused by neuritis. This assumption is based on the simultaneous appearance of other neural inflammations with these disturbances. In analogy with the findings from meningitis cerebrospinalis sections, COLLET concludes that mumps deafness could be the result of a serous meningitis. This author stresses the fact that it would be a mistake to assign too narrow a topography to this disease. In his opinion, a combination of meningeal ganglion lesions and neuritis are frequently present.

DE KLEIJN, 1944, in a very extensive study, described 21 patients, two of whom had a bilateral perceptive deafness. In his opinion, a disturbance of the central vascular system was the cause in these two patients, while the same cause was considered probable in the remaining 19 cases.

FUCHS and ALMOUR, 1947, assumed an allergy to be the origin in a patient who suddenly became deaf and who suffered from hayfever, bronchial asthma, migrain and sporadic attacks of dizziness. They got good results with injections of epinephrine and the elimination of allergens.

RASMUSSEN, 1949, published 18 cases of sudden perceptive deafness of which 15 also had disturbance of the vestibular organ. In his etiological discussion he considered a hemorrhage or thrombosis in the cochlea to be the most likely cause in 3 cases in which arteriosclerosis and hypertension were present. Vasospasm was considered in another case, and for the remaining cases a neuritis of the VIIIth nerve was suggested. He investigated the cerebrospinal fluid of 10 patients and for only one found an increased percentage of protein.

LINDSAY and ZUIDEMA, 1950, described 16 cases of sudden perceptive

deafness, of which 4 were accompanied by systemic diseases and 12 showed some degree of vestibular symptoms. In these 12 patients they considered the possibility of vasomotor disturbances of the inner ear and toxic conditions in the labyrinth.

According to FOWLER, 1950, in these types of deafness psychosomatic disturbances are responsible for contraction of blood vessels, sludging of the blood and congestion, resulting in anoxemia and an increased capillary permeability. Among his 26 patients he found that 18 had a history of psychic disequilibrium. He investigated the sludging phenomenon in 5 patients, but could only establish it once.

MYGIND, 1951, suggested a sudden increase in the intralabyrinthine pressure, and had good results in one patient with dehydration therapy. He further suggests that in cases which show no tendency to recover, dextran of macrodex may be used for dehydration purposes.

OPHEIM, 1951, who started with the assumption that an intralabyrinthine pressure rise would cause the deafness, performed a decompression operation on 3 patients. He saw only a slight improvement in one patient as a result of the operation. The operation only once disclosed signs of increased pressure.

KLOTZ, 1952, discussed the differential diagnosis of acute perceptive deafness. He listed psychogenic deafness, labyrinthal hydrops, vasospasm and cochlear hemorrhage as probable causes. As therapy he suggested vasodilatators and antihistamines. In cases of obstinate oedema he suggested decompression of the labyrinth, as described by OPHEIM.

SACHER, 1952, assumed a cochlear hemorrhage to be the causative factor in 5 patients with a sudden perceptive deafness. Four of these patients recovered completely. The only exception was a pregnant woman.

MOULONGUET and BOUCHE, 1952, saw 2 patients who had suddenly become deaf: the first, resulting from a serum injection, responded to nicotinic acid and antihistamines, and the second was a case of Ménière's disease caused by fat intolerance.

SALTZMAN and ERNSNER, 1952, and also VAN CANEGHEM, 1953, each described 3 cases in which it was assumed that a thrombosis of cochlear vessels could have been the cause, due to arteriosclerosis and advanced age.

In 1955, VAN DISHOECK and BIERMAN pointed out that sudden perceptive deafness is one of the symptoms of various diseases. In about half of their 17 patients, a virus infection was demonstrated serologically and suggested as the possible cause.

HELLER and LINDENBERG, 1955, described 5 patients for whom a virus infection was likewise suggested as a possible origin because they were all suffering from head colds.

HALLBERG, 1956, published 178 cases of sudden perceptive deafness seen at the Mayo Clinic. In 89, vascular disturbance was accepted as

the cause, and in 56 cases suffering from Ménière's disease it was thought to be an increase in labyrinth pressure. The remaining 17 patients could not be classified under either heading.

SISCHKA, 1956, described 5 patients who were cured after daily Ronicol infusions, (a nicotinic acid preparation). This therapy was continued until normal hearing was restored. Longstanding deafness did not respond to this therapy. The author thought that vasospasms were eliminated by improved blood supply, but also that the stimulation of the increased circulation of blood had a favourable effect on the diseased organ of Corti.

BOCCA and GIORDANO, 1956, published 28 cases in which the deafness appeared shortly after exposure to draft or cold, or during the course of influenza or a cold. They came to the conclusion that neuritis of the VIIIth nerve must have been the cause because no recruitment was found in 5 patients investigated and the caloric reactions were normal in 19. Agreeing with SVANE-KNUDSEN, 1957, who studied 21 patients suffering from sudden perceptive deafness, BOCCA and GIORDANO are of the opinion that the disease shows some analogy to rheumatic facial paralysis.

CLAVERIE, 1956, in a literature study of perceptive deafness occuring in the course of some well-known viral diseases, also described his personal observations. His results were the following: in rubella acquired postnatally and in influenza, deafness does not occur. In measles, chickenpox, epidemic encephalitis and poliomyelitis it is exceedingly rare. Conversely, in mumps and herpes zoster oticum deafness was frequently seen. In this author's opinion, deafness following mumps and herpes zoster probably was due to an affection of the organ of Corti and to meningoencephalitis. Claverie concluded that notwithstanding the probability of virus infection as a cause of perceptive deafness, this etiology is mostly very uncertain.

STERKERS, 1956, reviewed several factors which might be considered as the etiology behind sudden perceptive deafness. He listed:

1. Infections such as syphilis, herpes zoster oticum, mumps, rubella, malaria, typhoid fever and other infectious diseases which may cause encephalitis.
2. Intoxications such as streptomycine, dihydrostreptomycine, neomycine, alcohol, tabacco, lead, etc.
3. Tumors: acute unilateral deafness may be observed with a pontocerebellar tumor.
4. Traumata: Fractures of the labyrinth and also acoustic trauma.
5. Essential: According to the author this form of deafness may often be seen in young adults. It is accompanied by recruitment and may be caused by viral neuritis, central lesions, emboly, hemorrhage, oedema and vasospasm.

BROWN, 1957, described 34 cases of sudden deafness, including 3 patients who had mumps with parotitis. The deafness appeared at the beginning of this mumps infection in one of these patients, in the recovery phase in another one, and in the third 9 days after the patient had completely recovered. In the remaining 31 cases he was unable to find the cause of the loss of hearing.

II

ETIOLOGY AND PATHOGENESIS

The object of this investigation has been to study the etiology and pathogenesis of sudden perceptive deafness. The review of the literature as mentioned above shows that nothing conclusive is as yet known about the etiology of the disease. We find suggested as causative factors:

1. Thrombosis, vasospasm or hemorrhage.
2. Neuritis of the auditory nerve.
3. Systemic diseases, primarily those of the blood.
4. Inflammation of the labyrinth due to infection elsewhere in the body.
5. Emotional conditions.
6. Acute rise in pressure in the labyrinth.
7. Ménière's disease.
8. Allergy.

Etiology and pathogenesis of sudden perceptive deafness are frequently confused, and causes are often mentioned because they are known to create acute disturbances elsewhere in the body. One example of this is the analogy drawn between acute deafness and apoplexy, cerebral thrombosis, dysbasia and embolic processes such as in the *arteria centralis retinae*. It is very probable that these vascular disturbances are occasionally the responsible factors. We accepted this etiology in its application to older patients.

On the assumption that indications for such vasospasms and also for arteriosclerosis should be found in the blood vessels of the retina, 54 patients suffering from sudden perceptive deafness were examined at the Ophtalmological Clinic (Prof. M. C. COLENBRANDER, M.D.). Constricted retinal vessels caused by arteriosclerosis were found in only one case, that of a 51 year old man. However, in 2 patients changes in the fundus were found resulting from operated glaucoma. These two patients had not suffered from Ménière's disease, and so presumably no increase in labyrinthal pressure was present. Still it is a curious fact that among our 110 patients, two cases of glaucoma should be present.

If vasospasm due to arteriosclerosis or hypertension were an important factor, it could be expected that in a group of patients suffering from hemicrania, hypertension, dysbasia, etc. an appreciable number would have noticed changes in the acuity of their hearing. To clarify this point, 85 patients with these diseases were questioned. Their

answers showed that none of them had ever noticed a sudden decrease in their hearing ability.

On the other hand, 21 patients showing a very sudden onset of deafness which suggested vasospasm were given intravenous injections of 240 mg. aminophylline twice a day. In 12 of these patients slight to moderate improvement in hearing resulted, as established audiometrically ½ hour after the injections. These observations indicate that in such patients spasms may indeed have played a role in the pathogenesis.

The mechanism by which bloodsludging occurs in the blood vessels is considered by FOWLER Jr., 1957, to have great significance. This intravascular agglutination of the blood must be distinguished from extra-vascular coagulation (clotting). The latter occurs only after damage to or rupture of vascular tissues, the former in the absence of any rupture and often without any detectable damage to the neuro-vascular or other body tissues. Intravascular agglutination of the blood is, according to Fowler, a very common phenomenon and can be provoked by seemingly extremely slight stimulae. Even something so slight as a pinprick, hearing a shocking word, or a conscious or unconscious thought can induce sludging. In this author's opinion sludged blood in the vascular channels of the auditory apparatus is frequently involved in the causation of sudden, total, permanent deafness, sudden transient deafness and in many of the concomitant symptoms. In most of these cases of deafness of otherwise unknown origin, he found no other explanation tenable. The author also believes that a major factor involved in the genesis of otosclerosis in susceptible people is an intravascular agglutination of the blood occasioned by repeated or continuing so-called "stresses", „frustrations" and „emotional hypersensitivity".

In addition to vasospasm, the direct effect of a toxic substance on the nerve cells must be considered. In analogy with the paralyses produced in poliomyelitis, toxic oedema and toxic degeneration must also be taken into account. The oedema would then explain the sudden onset and the degeneration and the permanence of the loss of hearing. Such a pathogenesis has also been stipulated as probable in rheumatic facial paralysis. Bocca states that Coassolo's observations indicated that chilling of the *N. facialis* produced paralysis only when the experimental animal suffered from a virus infection. It was assumed that the cooling created the predisposition for a specific localization of the viral lesions. A similar predisposition is known to exist in poliomyelitis, where localization has appeared in a leg or arm which had recently been injected. It is difficult to demonstrate such a predisposition for the organ of Corti. It is true that a patient is occasionally seen who is convinced that he became deaf after the affected ear had been exposed to severe cold. One of our own cases was a 48 year old workman who, two weeks after he had had a cold, bicycled home from work with his right ear exposed to a sharp wind and cold rain. The following day

he was totally deaf in this ear. His tympanic membrane was normal and the deafness was of the inner ear type. Such cases are, however, rare and usually no such predisposition can be demonstrated.

In a recent study in pathological anatomy, Lindsay investigated the effect of ligating the local artery and vein of the cochlea. Both obstructions rapidly resulted in a destruction of the organ of Corti, the effect from the artery faster than that from the vein. These investigations indicate that vasospasm, embolism, and thrombosis can be important causes of degeneration of the cochlea.

The coagulation mechanism of the blood was determined in 15 patients. Only 3 had a slight decrease in tolerance of heparine and shortening of the prothrombine time. Consequently this mechanism can not be a major cause of the disease.

As mentioned above, many investigators searching for an etiological factor begin their reasoning on the basis of the striking similarity between a sudden increase in deafness, as seen during an attack of Ménière's disease, and a sudden attack of perceptive deafness, with which we are here concerned. They presume that despite the differences in the clinical picture increase of endolymphatic pressure is probably the cause of both diseases. This increase in pressure was established by HALLPIKE and CAIRNS, 1938. Since then this histopathological picture, consisting of a swelling of the ductus cochlearis without inflammation, has been frequently confirmed. As a result of these observations, the terms "endolymphatic hydrops" and "labyrinthine dropsy" are now used to designate this disease. The actual cause of the hydrops itself, however, remains unknown.

HILGER, 1950, attempted to show that the vital intralabyrinthine structures are dependent upon the adequate functioning of the end arteries which have no anastomoses there. Disturbances in this circulatory system, such as spasm, may cause such changes in the labyrinth as neuro-vascular ischemia and endolymphatic hypertension and result in Ménière's disease. It is quite possible that the same mechanism can produce acute deafness, either with or without hydrops.

WILLIAMS has suggested that Ménière's disease is a cholinergic hyperactivity. He goes on to say that this disease could be a form of intrinsic allergy in which the capillaries of the stria vascularis are involved, and that the disturbed endolymphatic pressure is a result of a localized extracellular oedema caused by the spastic-atonic condition of the capillaries. In accordance with this DAY, 1950, stated that some cases originate from a specific protein allergy. He also presented evidence that in labyrinthine hydrops the cochlea is affected first, and the vestibular disturbances follow secondarily. LEMPERT and co-workers, 1952, reported on Ménière patients who underwent partial labyrinthectomy. In them cystic changes on the inside of the membranous walls of the endolymphatic semicircular canals were found. They postulated that

such a Ménière's attack could be caused by the rupture of one or more vesicles with a resultant release of a toxic fluid in the endolymphatic ducts. They regarded Ménière's disease primarily as a condition of the vestibule and the pathological anatomic changes and symptoms in the cochlea as resulting therefrom. The cumulative deafness during the course of this disease was explained by a progressive destruction of the organ of Corti resulting from the repeated contamination of the endolymph from the vesicles.

However interesting these observations may be in respect to Ménière's disease, in our opinion they cannot be accepted as the basic cause of acute deafness, or perhaps only so in exceptional cases, because in acute deafness one of the most outstanding factors is the sudden damage in the cochlea with no previous or subsequent history.

SHIMANOTO and co-workers, 1954, reported a few interesting experiments on guinea pigs in which the endolymph was replaced by potassium chloride solutions in various concentrations. It was their opinion that disturbances in the secretion or absorption of the potassium were possible. Unequal high concentrations in the endolymphatic ducts might cause a syndrome of the vestibular apparatus resembling that of Ménière's disease. They also considered these high potassium concentrations responsible for the cochlear symptoms. Thus although little is known about it, we also think that the role played by electrolytes in the endolymph and perilymph must be taken into consideration in the etiology of acute deafness.

HALLBERG, 1956, found among 1270 Ménière's patients at the Mayo Clinic only 56 (4.4 %) with sudden deafness at the onset of the disease. We may thus conclude that sudden perceptive deafness rarely occurs in Ménière's disease. Vertigo attacks were reported by only 3 of our patients, and these cases were for this reason omitted. As stated, the isolated attack of sudden perceptive deafness forms a clinical picture quite different from that seen in Ménière's disease which is characterized by recurring attacks of true rotatory dizziness of short duration and the gradual development of basic deafness with transient worsening of the deafness during the attacks. Sudden perceptive deafness is, however, in actual cases accompanied by non-rotatory dizziness of longer duration.

Because of the fact that in Ménière's disease vertigo and sudden increase of deafness are both present, two separate conditions may be involved: hydrops or oedema of the semicircular canals causing rotatory dizziness and of the cochlea causing sudden deafness. For this reason, 4 patients who complained more than the others of dizziness were treated by dehydration therapy. The result was, however, not convincing.

Emotional factors are occasionally mentioned in the literature as causes of sudden perceptive deafness. FOWLER Jr. and ZECKEL, 1952-1953, stressed this etiologic factor. In our experience, however,

this psychosomatic influence appeared to be more the result of the deafness than its cause. Four of our patients felt that their deafness might result from their nervous tension and in only 5 others did we find definite emotional disturbance in the patients' histories. In only one case could a psychogenic deafness be established by means of the Stenger test and the "delayed speech" test. This patient was omitted from the study. In the remaining 8 patients a vasomotor disturbance of psychosomatic nature might have played a role.

There seems to be little evidence that allergy might constitute a significant cause. FUCHS and ALMOUR, 1947, described a patient in whom they presumed that allergens were responsible for a recurring hydrops of the cochlea resulting in sudden deafness. HELLER and LINDENBERG, 1955, also suggest allergy as a cause. Only 2 of our patients with acute deafness had a positive allergic history. Both patients were treated with Postafen and Sandosten and in one of them a definite improvement was audiometrically established. In our experience with about 3000 allergic patients per year there has been no instance of a case of acute deafness. In addition, allergy affects $\pm$ 10 % of The Netherlands' population, so that it is surprising that among 110 cases of acute deafness allergy is found so seldom.

In 1955 we suggested virus infection as a possible cause of sudden perceptive deafness.

The results obtained from our further study of this possibility are discussed below.

III

MATERIAL

The material studied consisted of 110 patients with sudden perceptive deafness of unknown origin. In the period between 1954 and September 1957, 74 patients were studied and treated in the Ear, Nose and Throat division of the University Hospital in Leyden. The data on the remaining 36 patients were obtained from Ear, Nose and Throat specialists in The Netherlands, to whom the clinic in Leyden is therefore much indebted.

Among these 110 patients, 68 were men and 42 women (see Table 1). The youngest patient was three years old and the oldest was 73. The average age of the group was 42 years. This average is roughly the equivalent of RASMUSSEN's, 1949, (that is, 8 under and 10 over the 40th year), but it is appreciably higher than that of FOWLER, 1950, (77 % under 40) and of LINDSAY and ZUIDEMA, 1950 (a majority was under 30 years of age).

TABLE 1

Distribution by age, sex and ears for 110 patients suffering from sudden perceptive deafness

Age in years	Men		Women		Total
	Unilateral	Bilateral	Unilateral	Bilateral	
0—10	1	4	3	1	9
10—20	—	—	4	—	4
20—30	7	2	4	1	14
30—40	12	2	8	—	22
40—50	11	4	8	—	23
50—60	12	1	6	1	20
60—70	11	—	3	1	15
70—80	1	—	—	2	3
Total	55	13	36	6	110

Men slightly outnumbered the women. The ratio of men to women was approximately 3 : 2. FOWLER, DE KLEYN and others got the same figures, while RASMUSSEN and SVANE-KNUDSEN report opposite results. The statistics of these authors, however, are so meager that they cannot be regarded as significant. Among our 110 patients the deafness was bilateral in 19 cases and unilateral in 91, or a ratio of $\pm$ 1 : 5. This

more or less agrees with the literature. In 44 patients the right ear was affected and in 47 the left. No preference for right or left ear could therefore be found, nor does the literature indicate any such preference.

No connection with any particular season of the year was found. An exception is formed by the Columbia-S.K. cases which were seen in the winter of 1954, as will be reported below. Very probably there was a small epidemic at that time.

IV

SYMPTOMATOLOGY

It is by now clear that a detailed history of the patient's illness is of the utmost importance in cases of sudden perceptive deafness. The symptoms appearing before, during or after this deafness are: fever, tinnitus, dizziness, headache, nausea, vomitting, and diplacusis. These symptoms warrant consideration here.

Deafness: While the onset of the deafness itself can be extremely sudden, as the literature also indicates: "a stroke or a bolt from the sky", it more frequently appears in the course of an hour or a day, and sometimes over a period of several days. Some patients first notice their deafness on awakening, others during use of the telephone, in bed when the good ear is on the pillow, or when directional hearing is disturbed. As mentioned before, the deafness can be either unilateral or bilateral. In general, an acute bilateral loss of hearing is noticed immediately, at the exact moment at which the hearing threshold becomes higher than the normal conversation level. This implies that the loss must be greater than 50 to 60 decibels. Discovery of unilateral deafness is frequently dependent upon how soon the patient's attention is directed to it or on some chance circumstance. In cases of sudden one-sided deafness with only a small hearing loss, it is very probably not noticed at all.

As an example of an acute and very sudden unilateral deafness, we can take the case of one of our patients who in the course of a telephone conversation suddenly heard nothing more. HOLMGREN, reports a case of an obstetrician who while doing routine auscultation of the foetal heart during a delivery, suddenly was unable to define the heart tones any longer. He concluded wrongly that the foetus had succumbed. It was later discovered that the ear which the obstetrician had been using for auscultation had suddenly become totally deaf.

An example of sudden bilateral deafness from our material is that of a mother who while waiting for her children heard the clock strike a quarter of eleven and failed to hear it at eleven. Another case is that of a stenographer who in the course of recording a parliamentary speech felt his hearing decrease until he was no longer able to hear anything and thus was unable to continue his work.

Fever. Twenty-nine patients were found to have or to have had fever. The importance of this symptom must be stressed, especially in relation to virus infections. As will be reported later on, in no less than 21 of

these patients was it possible to demonstrate the virus infection serologically. In 16 other patients who had severe headcolds at the onset of deafness but whose temperature was not taken and for whom serological tests could not be done or gave negative results, the possibility of a virus infection must still be considered.

Tinnitus. It occasionally happens that the tinnitus is so severe as to conceal the deafness. On the other hand, it is often the symptom which reveals the deafness by drawing the patient's attention to the ear. It was present in 78 patients. It preceded the deafness in 17 patients, which suggests that the disease was present before the deafness appeared. In only 5 patients did the tinnitus follow the deafness. In the remaining 56 patients it was noticed simultaneously with the loss of hearing. It is striking that in 32 patients the tinnitus, which is usually an indication of a cochlear lesion, was entirely absent. In these cases the lesions may have been supracochlear. Cochlear lesions, however, such as those caused by trauma, can appear independent of tinnitus.

In these types of sudden deafness, tinnitus is usually a temporary symptom. In approximately half of the patients it had disappeared within a month, and in the other half after a longer interval. In a few cases it appeared to be permanent.

Dizziness. The true rotatory vertigo which characterizes Ménière's disease rarely appears in these types of deafness and is surely not normal for them. This also applies to spontaneous nystagmus, which we saw in only 9 cases, one of whom was a patient with mumps meningitis, another a case of hypertension, and still another in whom the deafness seemed to be caused by vasospasm.

Dizziness was found in a total of 52 patients of whom 20 noticed it before the loss of hearing, 10 after it, and 22 simultaneously with it. In only two cases of vertigo did the symptom last more than a few months.

In general, dizziness can be expected to disappear within one week.

Headache. This symptom is often present in cases of accompanying virus infection. The localization of the headache is often in the neck, pointing to the possibility of a meningitis. Headache appeared in 24 patients, and in 10 of them it was an early symptom.

Nausea and vomitting. These symptoms appeared in only 14 patients, in 7 of them occurring rather. Usually neither is very severe, and as a result they are rarely mentioned spontaneously by a patient. They too point to a "benign" meningitis.

Diplacusis. This is a symptom usually only elicited by questioning of the patient. Like audiometrically-tested recruitment, it indicates a cochlear localization of the process. It was found in 73 patients. Four of these became aware of their deafness by this symptom, the others mentioned it on inquiry. Mostly the diplacusis appeared to be temporary. This symptom persisted in only 2 patients.

V

DIFFERENTIAL DIAGNOSIS

When a patient suddenly becomes deaf, this tragic event may be due to causes which vary widely. In the most favorable circumstances, deafness may be due to an impairment of the middle ear. Sudden deafness of the conductive type is not so rare. Even in otosclerosis, sudden deteriorations are known. In suppurative otitis media, sudden changes in hearing acuity depend on the secretion. This secretion may either improve or damage the hearing acuity. If the round window is protected from sound by a droplet, hearing improves; if the external meatus is filled with secretion, the hearing will be adversely affected. However, in such cases there is no doubt as to the middle-ear origin of the changes. By contrast, sudden deafness caused by catarrhal otitis media is often not recognized. In these cases the cavum tympani is partially filled with yellow fluid. The deafness is due to closing of the round window. Sometimes changing the position of the head is sufficient to restore hearing. The diagnosis of this disease can be made from the fluid-line seen through the tympanic membrane, from air bubbles passing through the fluid on insufflation through the Eustachian tube, and on measuring the tension of the membrane with the pneumophone. Moreover, the negative Rinne test, the lateralization of WEBER to the impaired ear, and the normal bone-conduction audiogram prove the existence of a conduction deafness. In these cases the puncture of the eardrum and the aspiration of the fluid will immediately restore close to normal hearing.

In sudden deafness of the perceptive type, the main problem is to indentify the origin, in order to make a differential diagnosis. If a trauma or purulent meningitis is present, the origin of the deafness is obvious. In the case of intoxication it may be more difficult. In the well-known deafness following quinine, salycilate and carbon monoxide, a possibility always remains that the deafness is wholly or partly due to the illness for which the drugs were taken. Moreover, carbon monoxide intoxication is often difficult to prove.

The localization of the process which caused the deafness can be of great value in diagnosing the etiology. For instance, if we can prove that the process is located only in the cochlea, the above-mentioned vascular mechanisms are probable, whereas if we can prove that the process is located in the nerve, nucleic area or auditory cortex, then a general toxic or virological cause is more probable.

The patient's hearing itself may help us, up to a certain point, to

make this differential diagnosis. To this end, the symptoms of recruitment, diplacusis and speech-discrimination loss are valuable. If these symptoms are present, a cochlear lession is very probable. It is of course possible to have combinations of cochlear and supracochlear deafness, and these are actually to be expected in acute deafness caused by virus infection or intoxication.

The differential diagnosis in psychogenic or simulated deafness may present considerable difficulties. In our experience and that of other authors, this problem is fortunately seldom met with in sudden perceptive deafness. For the moment, we have two satisfactory diagnostic means of exposing these patients: the Stenger test and the "delayed speech test".

As already mentioned above, and as we will see later, the differential diagnoses of sudden perceptive deafness and Ménière's disease are very important. In our opinion, these two diseases must be distinguished, notwithstanding the fact that in some cases the underlying cause may be the same.

VI

AUDIOLOGICAL INVESTIGATION

Seventy-four patients suffering from sudden perceptive deafness of unknown origin were observed audiologically. The tympanic membrane of the ear was normal in 61, scarred in 11 and perforated in 2 patients. In a very few cases the membrane in the deaf ear proved to be dull by comparison with the normal ear. The pneumophone value was determined as a matter of routine in those ears in which enough hearing remained. Only 15 patients showed a slight negative pressure, varying from - 6 to - 12 cm. water, and consequently a mild tubastenosis must be assumed in them. One of these patients showed a slight improvement in hearing after insufflation of the middle ear.

The audiometrical investigation was done as soon as possible and was frequently repeated during the illness and during the policlinic reëxaminations. Recruitment determinations, when possible, and the investigation of the vestibulary functions were done as often as possible. It must be kept in mind that in those patients who consulted us for the first time weeks or even months after the onset of their deafness, a spontaneous improvement may have taken place in the hearing as well as in the disfunction of the vestibulum.

In the 36 patients supplied by the questionnaire, only 13 had been tested with whisper voice and conversation speech. Audiometrical investigation and recruitment tests were not done on them.

The deafness was unilateral in 91 patients and bilateral in 19 patients Thus a total of 129 affected ears were investigated. The loss of hearing in the bilateral cases was practically symmetric in 12 and significantly disparate in 7 patients. (Table 2).

TABLE 2

Hearing loss in 19 patients suffering from bilateral sudden perceptive deafness. The combination of A and B was O, A and C was 1, B and C was found in 4 patients

Hearing loss	A: < 40 db	B: 40—80 db	C: > 80 db
Right ear	1	2	2
Left ear	—	2	3
Symmetric	2	6	6

The audiograms of this group of 110 patients had a wide variation in degree of deafness. A total loss of the hearing function was found in 57 patients (Tables 2 and 3).

TABLE 3

Hearing loss in 91 patients suffering from unilateral sudden perceptive deafness

< 40 db	40—80 db	> 80 db
4	41	46

The average loss among the total number of deaf ears was about 75 db (Fig. 1).

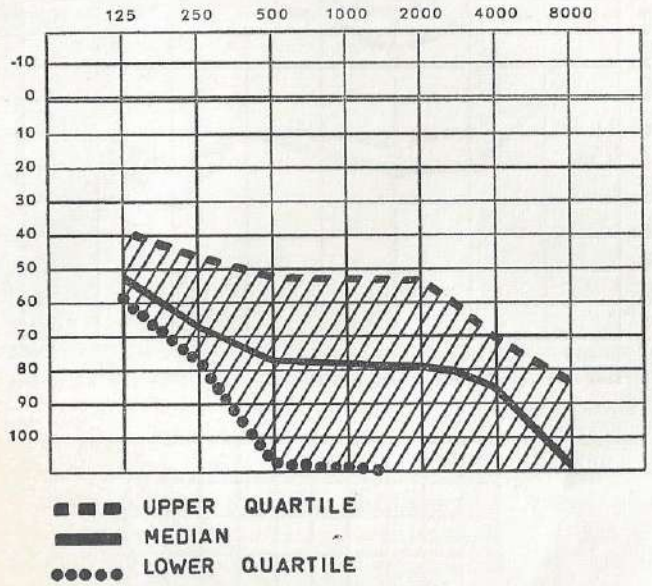
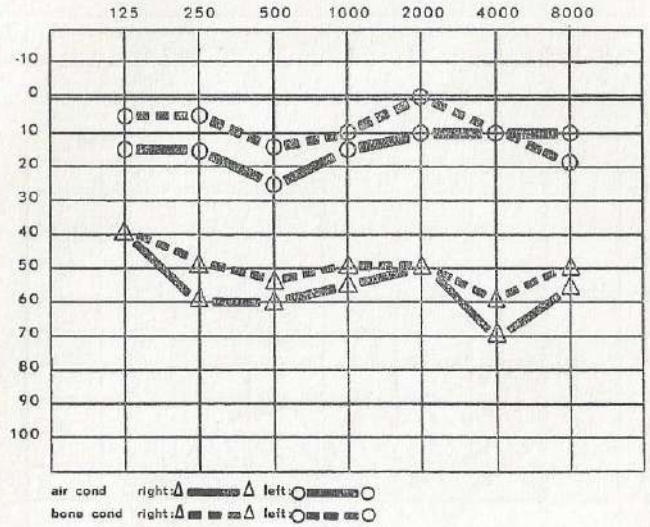


Figure I.
Median curve of audiograms of 110 patients suffering from sudden perceptive deafness.

Eighteen (16.4 %) patients with varying degrees of deafness had an approximately similar loss over the whole frequency range. An example typical of this group is case 1:

Case 1. One week before he visited the policlinic, a 25 year old medical student noticed on awakening that he was deaf in the right ear and troubled with ringing in this ear. One day later he was so affected by dizziness that he had to go to bed. The dizziness disappeared completely after 3 days, but the deafness and ringing persisted. He felt in addition very tired. The history showed no known contacts with infectious diseases and no intoxications. A perceptive deafness of the right ear was established audiometrically, presenting recruitment (Fig. II). The

routine E.N.T., general, neurological, and blood examinations showed no abnormalities. The pneumophone values were normal. The syphilis reactions were negative. The vestibule reactions were normal on both sides. The serological tests for virus were negative. The patient was treated with nicotinic acid and bed rest. Examinations after 2 and 4 weeks showed substantial improvement of his hearing.



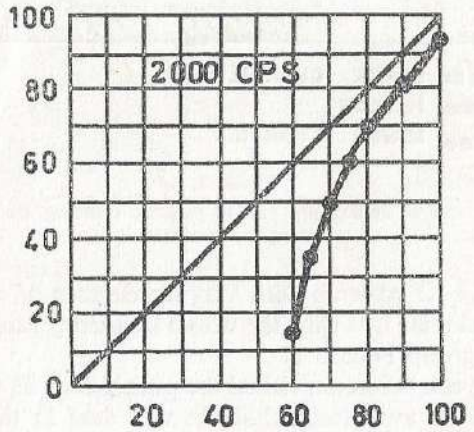
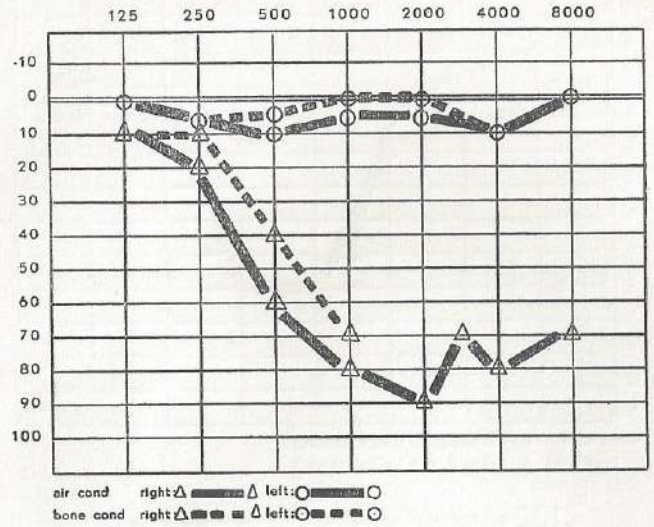


Fig. II.
Audiogram and results of the binaural loudness balance test in a patient with approximately equal hearing loss at all frequencies.

A loss of hearing primarily in the high frequencies, but of varying degrees, was seen in 22 patients (20 %). Case 2 illustrates this group.

Case 2. A 40 year old woman, after a week of intermittent ringing in the right ear, suddenly became deaf in that ear. A year before, she had been treated for a catarrhal otitis media of both ears. Ever, since her youth she had complained of a running nose and frequent head colds.



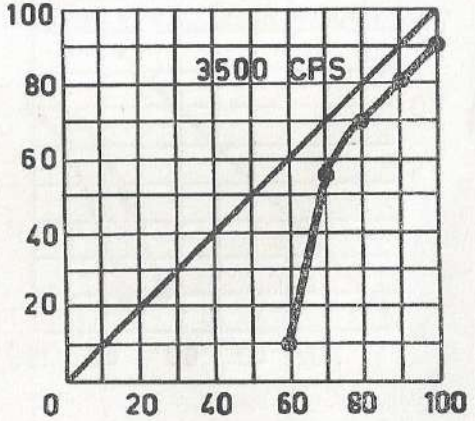


Fig. III.
Audiogram and results of the recruitment determination according to Fowler, in a patient with unilateral hearing loss primarily in the high frequencies.

For the past 10 years she had been bothered by asthmatic attacks. No dizziness or headache accompanied the onset of the deafness, and she was otherwise in good health. There was no history of contact with infectious diseases. Except for a rather swollen, moist nasal mucous

membrane, nothing unusual was found. Audiometrically she had on the right side an acute perceptive deafness with recruitment (Fig. III).

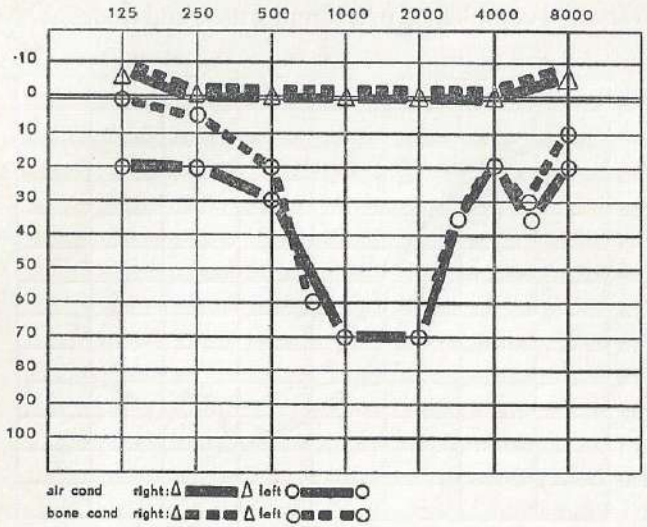


Fig. IV.
Audiogram and loudness balance test in a patient with a hearing loss in the middle frequencies.

The vestibulary, neurological, general, x-ray and allergic investigations showed nothing suggestive. Syphilis reactions were negative and the cerebro-spinal fluid was normal. The serological virus investigation was also negative. The patient received 240 mg. euphylline twice a day for a week, as a result of which the hearing improved daily. Because of the tendency to allergy in the history, she then received Sandosten

for 10 days. A reëxamination after 3 weeks showed that there was still an audiometric loss of 15 db in the middle frequencies.

A loss in the middle frequencies was shown by 7 patients (6.4 %). Case 3 demonstrates such a loss.

Case 3. A 40 year old man suddenly became deaf in the left ear after a week of ringing in that ear. In this period he had intermittent headache and became dizzy a day after the deafness appeared. He himself ascribed his deafness to having sat in a draft. In addition he was always much troubled by colds, sore throats and hoarseness. There had been no infectious diseases or intoxication. Examination showed the tympanic membranes to be somewhat dull, and the nose and rhinopharynx mucous membranes were rather red. The pneumophone values were normal. Audiometry showed a combined deafness with recruitment (Fig. IV). The investigation of the vestibular organ showed an entirely normally-reacting labyrinth. Neurological, general and x-ray examinations were negative. Syphilis reactions and serological tests were negative. As therapy, nicotinic acid was given by mouth for 8 weeks. Audiometrical reëxamination after 8 weeks showed a marked improvement, especially of the bone conduction.

Recruitment. As stated above, perceptive deafness can be the result of a cochlear or supra-cochlear lesion. It is frequently difficult to determine, especially in cases of total deafness, what localization is involved. A combination of these two possible localizations can also be present. The main differential diagnosticum for assuming the presence of a cochlear lesion is the symptom of recruitment.

It was FOWLER, 1941, who first associated this symptom with perceptive deafness. He had found in a patient with one-sided perceptive deafness that for a certain sound intensity the loudness sensation in both ears became equal. Such a patient with one normal ear and one deaf ear may have at the threshold a hearing loss, for instance, of 40 decibels, whereas at an intensity level of 70 decibels the loudness sensation is the same for the deaf ear as the normal one. We now know that recruitment is associated with cochlear lesions because DIX, HALPIKE and HOOD, 1948, in a series of investigations, were able to show that recruitment was absent in the majority of types of deafness resulting from *acousticus neurinomen.* In Ménière's disease, by contrast, this system was regularly present. As Ménière's syndrome is a purely cochlear affection, they concluded that the recruitment is a result of a cochlear involvement. Support for this lies in the fact that this symptom is also always present in acoustic trauma. That acoustic trauma results in cochlear deafness is shown by animal experiments in which cochlear degeneration, especially in the auditory hair cells, is always found.

At present it is possible to determine this symptom by the direct methods such as the binaural and monaural loudness balance test and the dynamogram; and by the indirect methods such as speech audiometry, adaptation tests, difference limen and self-registering threshold audiometry. Through a critical study made by TAUNO PALVA, 1957, we now know that recruitment determination in the technically possible cases can best be made by use of the direct methods. In cases where both ears have indentical or practically indentical thresholds, the most reliable results were obtained by means of speech and self-registering threshold audiometry. One very simple technique for studying the loudness function of the ear is that of Moll's dynamogram. However, in a study using 11 patients suffering from ponto-cerebral tumor, Rol and Bierman investigated whether this fact, which had meanwhile been published by Hallpike, could be ascertained by means of the dynamogram. It was confirmed that in these patients the binaural loudness balance test showed no recruitment. However, the dynamogram indicated a moderately steep curve. Consequently, extreme care must be exercised in drawing conclusions from a steepness in the dynamogram, only a very steep curve being significant.

Recruitment was investigated in 51 of our patients, of whom 9 had bilateral deafness. The existence of a cochlear lesion was assumed in 29 (56.8 %) on the grounds of an established recruitment. It is extremely likely that a supracochlear condition was present in the 22 cases who showed no recruitment. Because no neurological evidence was found in any of these patients, no certainty about whether the process was located in the auditory nerve or in the auditory centre was obtainable.

It is doubtful that vascular disfunction plays a great part in these cases, although there is opinion to the contrary. There would seem to be more chance of a toxic degenerative process or oedema such as is seen in neuritis and encephalitis. In cases of bilateral deafness also, a widely affective toxic agent is more probable than a hemorrhage or vasospasm in the cochlea.

VII

INVESTIGATION OF THE VESTIBULAR ORGAN

This investigation comprised 71 patients. Thirty-four of this group experienced dizziness at the onset of the deafness. The deafness was unilateral in 61 cases and bilateral in 10. Spontaneous nystagmus was only seen in 9 patients suffering from unilateral deafness. The caloric reactions, using 10 cc. of water at 30° and 44°, were determined at the first examination (Table 4).

TABLE 4

Vestibular reactions in 71 patients suffering from perceptive deafness

Deaf ear	1st examination within one month after onset	1st examination 1 month after onset or later	Total
Not sensitive	5	7	12
Reduced sensitivity	10	5	15
Hypersensitive	1	—	1
Preponderance	5	—	5
Normal	23	15	38
Total	44	27	71

Thus 33 of the 71 patients had deviations pointing to a disturbed functioning of the labyrinth. The number of lesions present in the group examined shortly after the onset of the deafness and those seen later was practically the same.

Among the 10 patients with a bilateral deafness, 5 had bilateral and 2 only unilateral disturbed function of the labyrinth. It is remarkable that the remaining 3 patients with subtotal deafness on both sides had normally reacting labyrinths.

In 26 patients it was possible to establish the simultaneous presence of vestibulary lesions and dizziness. At the same time there appeared to be a parallel between the seriousness of the lesions and the degree of deafness. The fact that none of these patients had a true vestibular rotatory dizziness and that only 9 showed a spontaneous nystagmus may, in spite of the presense of the vestibulary lesions, indicate a supracochlear involvement in these types of deafness.

VIII

VIRUS INFECTIONS

In February 1954, two patients were admitted to the clinic with a sudden perceptive deafness accompanied by signs of meningeal irritation and fever. Blood counts revealed only a relative lymphocytosis. Examination of the cerebrospinal fluid confirmed the suspicion of meningeal irritation (table 7, 1 and 2). Physical examination revealed no other neurological signs. These findings suggested a virus infection. With this in mind, blood, cerebrospinal fluid and faeces were investigated. The results were most surprising. In the faeces and cerebrospinal fluid of both patients an encephalomyelitis virus belonging to the Columbia S.K. group was isolated. The serum showed a marked increase of neutralizing antibodies for this virus (neutralization indices of 2000 and 5000). In addition, the serological examination also showed an insignificant increase in antibodies for mumps virus. It was not possible to determine whether this limited increase in these two patients was due to a recent exposure to mumps virus or to an infection by mumps virus long previous.

Encouraged by these virus-isolations and consideration that virus infections might play an important part in these perceptive types of deafness, a number of these patients were studied in this way.

This virus investigation was restricted to mumps virus, the influenza viruses and the "Enteric group" of viruses. For mumps virus, the American mumps strain (Enders) and the Dutch strain (GUNST) were used. The reactions were done with V(irus); S(oluble) and with unpurified antigens.

For the influenza investigation, antigens prepared from the A group (strain W.S. and P.R.S.), A^1 group (Liverpool and Stockholm) and B group (LEE and later BONN), were used. With all these antigens, complement-fixation reactions of the sera under study were performed.

For the investigation of the "Enteric group" of viruses, preparations were primarily made from faeces samples and spinal fluid in order to isolate Columbia S.K. virus, poliomyelitis virus or Coxsackie viruses. Either hemagglutination inhibition tests or neutralization reactions, or both, were performed with the sera.

Seventy-four of our patients were examined for the above-named viruses within 3 months after onset of their disease. In 66 cases in this group, serological reactions were done and investigation of faeces samples were carried out. In 8 cases, only serological reactions were done.

In addition, the cerebrospinal fluid of 49 patients was examined chemically and morphologically and searched for viruses.

A virus infection could be established in 25 out of 74 patients. In 4 cases a virus was isolated. The results were 18 cases of mumps, 7 cases of infection by the influenza group, and the "Enteric group" resulted in 7 cases, 5 of which were Columbia S.K. virus and 2 were Coxsackie virus (Table 5).

The fact that a few patients in whom the serological results were negative showed a "benign" meningitis and also that other patients had a grippe-like picture with headache and fever at the onset of the deafness, suggests other as yet unrecognized virus infections as the cause of this sudden deafness.

TABLE 5

Results of the various investigations of possible virus infections

Virus	Strain	Culture		Blood antibodies		Spinal fluid culture	
		pos.	neg.	pos.	neg.	pos.	neg.
Mumps	Gunst-Enders	—	—	18	56	—	—
Influenza	Type A en A^1	—	—	2	72	—	—
	Type B	—	—	5	69	—	—
"Enteric group"	Poliomyelitis	0	66	—	—	0	49
	Columbia S.K.	2	64	5	69	2	47
	Coxsackie	2	64	—	—	0	49

—: not investigated.

Thus a total of 32 positive results were found out of a total of 25 patients. Combinations of these positive results were found as follows:

a. Five cases of Columbia S.K. infection included 3 patients with insignificant rise in the antibodies for mumps virus, and in one a similar rise against mumps and influenza-B viruses was observed. It can thus be assumed that neither the mumps virus nor the influenza virus can be considered to be causative agents.

Inevitably, the question arose whether there is a possible relationship between Columbia S.K. and the mumps virus. This possibility was excluded by tests done by VERLINDE and WOLFF using cross-neutralization techniques.

It is, however, very likely that the slight rise in the mumps antibodies in these cases of Columbia S.K. infection indicates that a

hypersensitive condition of the reticulo-endothelial system is produced which releases other antibodies into the bloodstream. It should also be pointed out, in addition, that the Columbia S.K. virus, actually an enterotropic virus, is certain to create such a situation. We thus consider that this dubious increase of mumps antibodies must be taken as a non-specific reaction. These cases were therefore admitted as pure Columbia S.K. infections.

b. Among the mumps cases (18), one patient also showed an increase in influenza B antibodies. Also 4 as described under *a.*

c. The influenza cases (type A: 2 patients, and type B: 5 patients): Type B was demonstrated twice as indicated under *a* en *b.* One influenza type A case was found in a patient in whom a ponto-cerebellar tumor was later diagnosed.

Influenza virus:

Whether the influenza virus must be regarded as a causative factor in these types of deafness is still open to much doubt. Considering that influenza appears each year and that the antibodies certainly circulate in the body for three months, it could be expected that in any random group of 100 persons at least 7 would be encountered who had suffered from a serologically ascertained influenza. It is remarkable that during the last influenza A epidemic (1957) no cases of sudden perceptive deafness came to our knowledge. This can to some extent serve as confirmation of the above material.

Mumps virus:

Deafness in the course of parotitis epidemica is relatively rare but nevertheless very well known. The literature offers some information as to the incidence of deafness as a result of mumps. In 1944 McGuinness and Gall observed 1378 patients during a mumps epidemic in an army camp: they were unable to demonstrate a single case of associated deafness. Eagles, 1947, was also unable to establish deafness after observing 1664 mumps patients during a four years' study in an army camp. Laurance and McGavin, 1948, reported only one case of unilateral deafness among 203 cases of mumps. Despite this, it has been estimated by Scott and Crawford, 1949, that 3 to 5 percent of the cases of acquired total deafness in the United States are due to mumps; however, in all cases in reported epidemics it occured in only a fraction of one percent. Amarante, 1957, also presented some data on the frequency of mumps deafness. As early as 1802, Hintze first directed attention to this type of deafness in a man suffering from an epidemic parotitis. Toynbee in his book on diseases of the ear, described this complication in mumps patients. He asserted that the toxin produced during this disease is frequently the cause of total deafness and that the nervous system is undoubtedly damaged by it.

He saw the deafness frequently unilateral and sudden in onset. Mayer stated that deafness as a complication in epidemic parotitis occurs more frequently than generally recognized. Here he disagreed with Bezold, who found that only 1.7 % of the severe perceptive deafness cases are caused by this disease.

Conversely, Hubbard found among 50,000 seriously deaf individuals in the United States ± 3—5 % resulting from *parotitis epidemica.* According to him it was more frequent among men than women. In general it is considered that 65 % of the cases of epidemic mumps are followed by a lymphocytic infiltration from the meninges. David de Sanson and others described cases of meningeal reactions during this disease. De Sanson also reports a patient who had without question suffered an infection as a result of contact with the mumps virus and who showed meningeal symptoms and not those of parotitis. Since very few cases ever come to autopsy, the true nature of the pathogenesis is not entirely known. Authors such as Wesselhoeft and Brown state that a "relatively" high percentage of mumps cases may show changes in the spinal fluid without any clinical symptoms of meningitis. They further stated that the spinal fluid changes are most frequently present between the eighth and twelfth days of this desease.

According to Amarante the origin of the deafness in this disease must be sought in a toxic neuritis. He commonly saw this deafness in a unilateral form. His cases were primarily children in whom the localization of the condition could not be satisfactorily investigated. Coombs, 1952, also mentioned that the frequency of nerve deafness in mumps, encephalitis or meningeo-encephalitis varies considerably. Several other authors: Arey, Eberlein, Birnberg, Wallerstein and Bristow, agreed with this statement. Thus Coombs suggested that probably many cases of nerve deafness in children, especially the unilateral cases of unknown etiology, may be due to mumps, and our study gives proof of this supposition.

Mumps infections and reinfections without parotitis symptoms are very common. This fact is not generally known. Thus among our 14 reported patients who showed a serological mumps reaction, only one had parotitis. From this we can conclude that mumps infections usually lead to parotitis without deafness, or in exceptional cases to parotitis with deafness, and once in a while to deafness alone. These mumps infections leading to deafness without parotitis have been proved to be reinfections in persons who had contact with mumps patients. It has been established by Wolff, who in an extensive study carried out complement-fixation reactions for one large group of normal adults and another group without clinical symptoms after recent contact with mumps, that in the first group positive mumps reactions were extremely rare. Nearly all persons in the second group showed positive reactions for mumps without suffering from parotitis and even without being ill.

We have compared these results with the serological mumps reactions of our 74 patients suffering from sudden perceptive deafness (Table 6).

TABLE 6

Serological reactions for mumps in 3 groups. A mumps infection was assumed if the complement-fixation titers were 288 or more or became significantly higher or lower in the course of a month.

Material	Number	Positive	Negative
Normal Persons	857 (100 %)	1 (0.1 %)	856 (99.9 %)
Contact Persons	300 (100 %)	298 (99.3 %)	2 (0.7 %)
Sudden perceptive deafness	74 (100 %)	14 (18.9 %)	60 (81.1 %)

Table 6 indicates that a positive reaction occurs only sporadically in normal individuals, while it can almost always be found in contact individuals. The high percentage of acute deafness here is immediately striking. It is noteworthy that 16 of these patients had already had an earlier mumps infection. In all probability these reinfections must be considered as important causes of the disease. Case 4 can serve as an example of reinfection with mumps virus.

Case 4. A 42 year old man became suddenly deaf in the right ear about 2 months before the first examination. A week previous he had felt ill, but had been without fever. He was then troubled by ringing in the right ear, dizziness, nausea and vomitting. At the end of that week he noticed that everything sounded strange in his right ear, and one day later the deafness appeared. He had had no previous ear complaints. A month before the deafness appeared, his children had had mumps. He himself had already suffered from mumps a long time before. Three days after the onset of deafness, the dizziness, nausea and vomitting disappeared.

Routine examination disclosed nothing unusual in the ear drums, nose or throat. Audiometry showed a total loss of air and bone conduction in the right ear while the left ear showed a moderate perceptive deafness (see fig. V). Recruitment determinations were impossible because of the total deafness. The caloric testing gave normal values for both labyrinths. Nothing exceptional was found in the fundus of the eye or in neurological and general examinations. X-rays of the ossa petrosa (according to STENVERS and SCHÜLLER) were normal. Serological virus investigation for mumps, influenza and the "Enteric group" gave titers which indicated a previous mumps-virus infection. This confirmed the diagnosis of sudden perceptive deafness resulting from a mumps-virus reinfection. Administration of 100,000 units of vitamin A and Ac. nicotinicum 100 mg. three times daily by mouth showed no effect when checked audiometrically after 4 and 8 weeks. On the other hand, the

ringing in the ears was sharply reduced following this therapy, so that the patient was no longer handicapped by it.

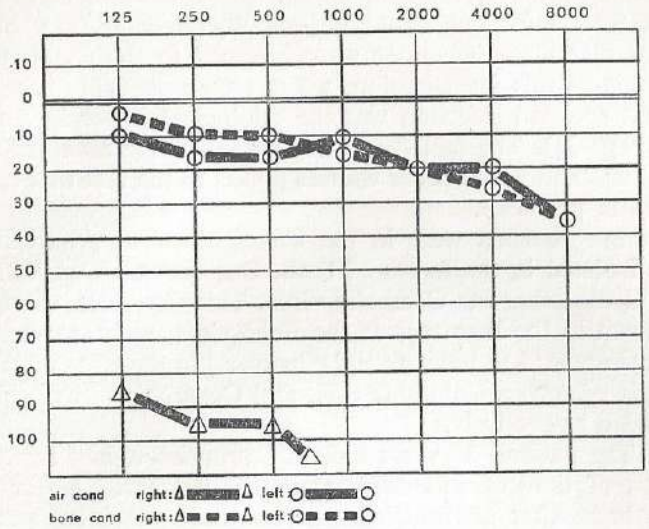


Fig. V.

Typical audiogram of a patient suffering from mumps reinfection.

There are varying theories about deafness resulting from a mumps-virus infection. The most obvious is that it is caused by toxic neuritis, although this has not yet been clearly demonstrated. It was only possible to investigate the recruitment phenomenon in 6 out of 14 patients presenting positive mumps reactions. It is striking that recruitment was present in 4 of these patients indicating that in them a cochlear lesion must be present. In addition, neuritis of the auditory nerve and encephalitis are also very likely to be present in some patients.

"Enteric group":

It is known that both the poliomyelitis virus and the Coxsackie virus can be pathogenic for man. Until recently this has not been generally accepted for the Columbia S.K. virus. Like the poliomyelitis virus, the Columbia S.K. virus gives a poorly differentiated clinical picture. Such symptoms as fever, with or without mild gastro-intestinal disturbances, angina, headache, muscle pain and possibly meningeal symptoms can appear. Evidence that Columbia S.K. infection probably exists in The Netherlands is given by the presence of neutralizing antibodies in the gammaglobulins of blood donors. These antibodies have also been found in the serum of patients with encephalomeningitis, encephalomyocarditis, aseptic meningitis or a febris e causa ignota.

VAN TONGEREN and VERLINDE, 1954, were able in an extensive

investigation to establish the pathogenesis for man by isolating this virus in faeces, throat mucous and spinal fluid. Only some separated cases were found and no epidemiological connection between these Columbia S.K. infections could be demonstrated. It was, however, determined that these infections were confined to the winter months. On the basis of this investigation, we can very probably accept it as certain that such an infection was the etiological factor in 5 patients (see also page 31). The fact that these patients were seen only in the winter of 1954 and not in other winters points to the fact that epidemic connection is probable.

Two of our patients were in the Coxsackie virus group and this virus was isolated in the faeces of both. One showed Coxsackie virus type A and the other the Coxsecho virus. This Coxsecho virus, which is still known in the literature as the Echo-virus, must, in view of its behaviour in regard to baby mice, be included in the Coxsackie group. An example of such a Columbia S.K. and Coxsackie virus is given by cases 5 and 6 respectively.

Case 5. The patient, 39 years old, café proprietor, had for 5 weeks complained of tinnitus and deafness in the left ear. This developed within an hour after his noticing that everything sounded strange in that ear. A few days before the onset of these complaints he had had a slight cold, but his hearing was still good. During the preceding winter months he had had other colds. He smoked about 30 cigarettes a day, usually drank moderately but occasionally took as many as 20 glasses of beer a day. He had had no venereal or infectious diseases and no recent contact with virus infections was known.

Routine ear, nose and throat examination showed a normal appearance of the tympanic membrane. The mucous membranes of nose, throat, fauces and rhinopharynx were hyperemic. Sinus x-rays were normal. The audiogram showed a perceptive deafness on the left side (fig. VI*a*). Recruitment could be demonstrated by both dynamogram and speech audiometry. Examination of the vestibule showed no spontaneous nystagmus, and caloric tests gave normal values. Neurological, general, x-ray and opthalmological investigation were equally negative. Hematologically there was a relative lymphocytosis, with a slightly increased leucocyte count. Blood sedimentation rate was normal. The spinal fluid, by contrast, showed clear indications of a meningeal irritation (Table 7: No. 1). Virological tests were positive for Columbia S.K. and questionable for mumps. Columbia S.K. virus was grown from faeces, spinal fluid and throat mucous and there was at the same time a markedly accelerated formation of neutralizing antibodies in the blood against this isolated virus (neutralization-index: 5012). It was a striking fact that the patient had no fever during the observation period. We attempted to influence this picture therapeutically with 2 grams of aureomycin daily for 5 days combined with

vitamin B_1 and B_{12}. The result with respect to the ear was nil, however. About 6 weeks after the onset of the deafness on the left side, a sudden impairment of hearing appeared in the right ear (fig. VI*b*). Recruitment was also present in this ear. Repetition of the examination of the vestibular organ showed normally sensitive labyrinths. Novocaine injections of 10 cc twice a week for 8 weeks, were then used to attempt to improve this condition, but audiometric reëxaminations after 2, 3 and 6 months showed absolutely no effect.

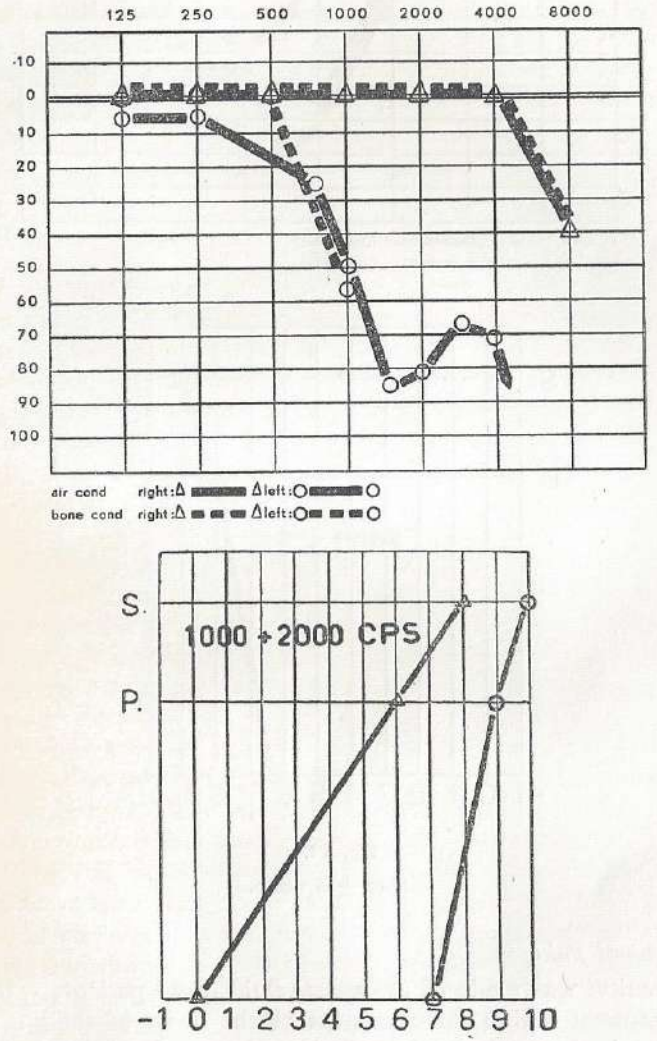
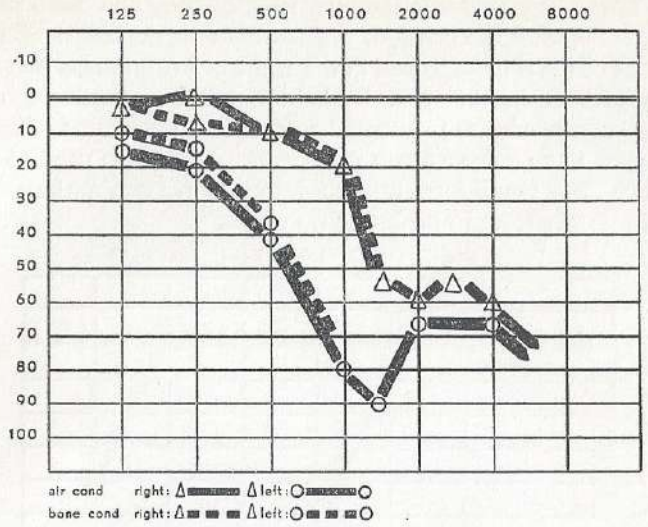


Fig. VI.
Audiograms and dynamograms of a patient suffering from a Columbia S.K. virus infection. *a.* 1st examination



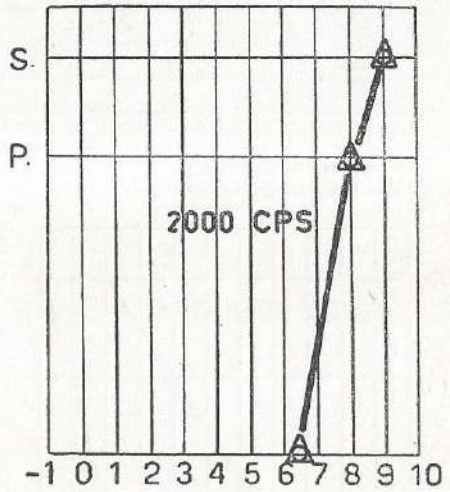


Fig. VI.
b. after ± 6 weeks.

Cerebro-spinal fluid

Examination was made of the spinal fluid of 49 patients who came under treatment within three months of the onset of their deafness. An increase in the total protein, a limited pleocytosis, a positive NONNE and/or PANDY test, and a disturbed benzoin and/or gold-sol test was seen in 14 of them (28.6 %).

From Table 7, it can be seen that in 8 of the cases with meningeal irritation a virus infection could be determined. The two mumps cases (nos. 6 and 7) had no parotitis. In the cases in which we were unable to demonstrate a virus there is nonetheless a substantial probability that this was still the case; the picture of a "benign" meningitis was present. We have therefore included these patients in the virus group. It must also be considered that if all these patients had been examined sooner after the onset of the deafness the number of cases of "benign" meningitis would undoubtedly have been larger.

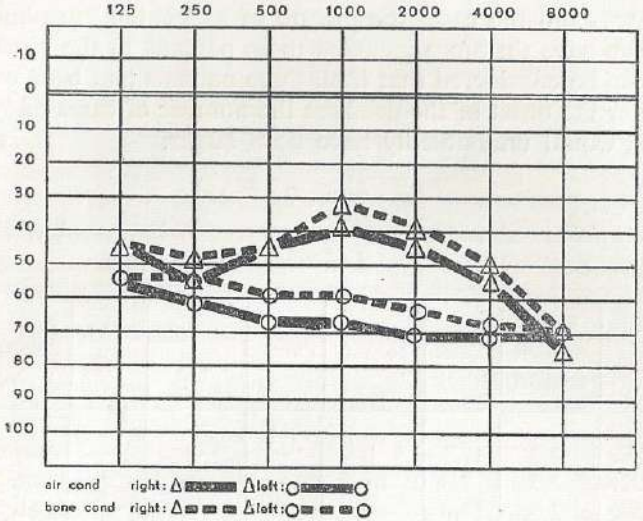
TABLE 7

Cerebro-spinal fluid examination for 14 patients seen within 1 week of the onset of deafness.

No.	Etiological factor	Total protien content	Cells	Nonne	Pandy	Benzoin test	Gold-sol	Pressure	Queckenstedt
1.	Col. S.K.	2,2	17/3	+	+	→	—	norm.	+
2.	Col. S.K.	2,6	7/3	sp.	+	→	↑	,,	+
3.	Col. S.K.	1,0	17/3	sp.	sp.	—	—	,,	+
4.	Col. S.K.	1,3	8/3	—	sp.	→	↑	,,	+
5.	Infl. B.	1,7	4/3	—	sp.	→	—	,,	+
6.	Mumps	1,0	7/3	sp.	+	→	—	,,	+
7.	Mumps	1,4	7/3	sp.	+	→	—	,,	+
8.	Coxsackie	1,9	11/3	sp.	+	—	—	,,	+
9.	Unknown	1,0	22/3	—	sp.	→	↑	,,	+
10.	Unknown	1,1	10/3	—	+	→	↑	,,	+
11.	Unknown	1,5	32/3	—	+	→	↑	,,	+
12.	Unknown	2,1	7/3	sp.	+	→	—	,,	+
13.	Unknown	1,2	12/3	—	—	→	—	,,	+
14.	Unknown	1,4	6/3	sp.	+	→	—	,,	+

Case 6. A 38 year old man had had a cold for about three days and had a continuous slight pain at the back of his head. On the third day of the illness he suddenly became deaf in both ears. The hearing became continually worse and after 14 days he was almost completely deaf. The patient was also dizzy and had a loud ringing in his ears. The day before the onset of deafness everything sounded strange. After a few days in bed the dizziness disappeared. The hearing had then improved slightly but was very variable. The ringing remained constant. Two months after the onset of the deafness he was admitted to a hospital where a diagnosis of lymphocytic meningitis was made on the basis of a lumbal puncture. The hearing complaint was treated with pilocarpine which was at first successful. After two weeks he had a relapse and

was then again practically deaf. Nicotinic acid administered intravenously then gave some improvement and the ringing was reduced. A month later he had trouble with a sinusitis maxillaris on the left side



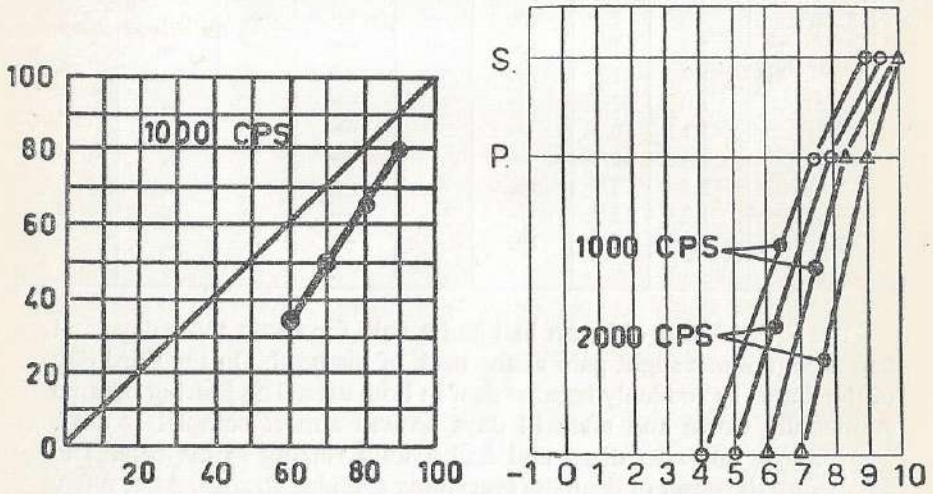


Fig. VII.
Audiogram and binaural loudness balance test in a patient with a Coxsackie virus infection.

which cleared up after a week of conservative therapy. Two months later the deafness again became worse and the ringing reappeared. He was then referred to us and admitted to our clinic for examination.

Except for somewha tdull tympanic membranes nothing was found in routine examination. The pneumophone test gave for both sides a pressure of - 8 water. The audiogram showed on both sides a perceptive deafness, with a greater loss of hearing on the left side. The recruitment determination was negative (fig. VII) so that a supracochlear involvement was suspected. Neurological examination showed a slight deviation to the left in the walking and past-pointing tests. The results of the electroencephalographic examination was a mildly diffuse, unspecific, primarily left tempero-parietal. Examination of the vestibule showed first a nystagmus preponderance to the right and a few days later to the left. There was also a positional nystagmus to the right when lying on the left side and toward the left when lying on the right side. The opthalmological and hematological investigations were negative, as were the syphilis reactions. The spinal fluid, faeces and blood were examined for virus. The result was the isolation of Coxsackie virus (Echo 9) from the faeces. We thus may diagnose a mild meningo-encephalitis, very probably caused by a Coxsackie virus infection. Therapeutic treatment with aureomycine for 6 days had no result. At the same time the patient received daily intravenous injections of aminophylline, which resulted in a slight improvement in hearing that could be audiometrically determined within half an hour. After 5 weeks of observation he was discharged, without any permanent improvement. It was noteworthy that this patient, at a threshold of 60 db right and 80 db left, reached a speech audibility of 85 % with sufficient amplification.

IX

CLASSIFICATION OF THE PATIENTS

Arrangement of these 110 patients according to possible etiology of the deafness was based on the facts and considerations mentioned above. Virus infection as a cause of sudden perceptive deafness is undeniable whatever the pathogenesis may be. This etiology was not only accepted on the basis of positive serological reactions but also when a "benign" menigitis or severe grippe of the head at the beginning of the disease was reported. The presence of vasospasm in virus infections was supposed in 7 patients out of a group of 16 who were treated successfully with spasmolytics.

Arteriosclerosis was assumed to be the etiology of the sudden deafness in 12 older patients who also showed other localizations of the disease. In this affection the pathogenesis can just as easily be vasospasm or thrombosis as bleeding. The possibility of spasm was considered when satisfactory results were obtained with spasmolitics. In the cases where a very sudden onset took place without any later improvement, the possibility of a hemorrhage or thrombosis was more likely, although only 3 patients had a very slight change in the coagulation mechanism, in the sense of a slightly lowered heparine tolerance and reduction of the prothrombine time.

In spite of the absence of vertigo, a genuine hydrops was considered in 4 patients. The dehydration therapy, however, gave no convincing result, so that our conjecture here had no actual support.

Emotional factors were present in 8 patients. Use of the Stenger test and the "delayed speech test", however, showed that in these cases the deafness was not psychogenic. It is possible that this type of deafness originates in a psychosomatic mechanism by which the emotion causes vasospasm in the cochlea. This cannot of course be demonstrated, although dilatation and contraction of the blood vessels, for example in blushing, is a very common mechanism.

Only 6 patients contracted their sudden deafness in the course of another disease, one after an appendectomy under general anesthesia, two as an early symptom of ponto-cerebellar tumor, one with BESNIER-BOECK disease, one with a positive WASSERMAN reaction and another suffering from a multiple sclerosis. Some of these diseases are known to be causes of perceptive deafness, others not. However it is not usual in such cases for the deafness to start suddenly. In 15 patients no etiological factor of any sort could be found.

Undoubtedly unrecognized virus infections with no marked symptoms

TABLE 8

Classification of the probable etiology in 110 patients suffering from sudden perceptive deafness

Etiology		Number	Localization		Sex		Deafness			Recruitment			Tinnitus	Diplacusis	Dizziness	Vestibular lesion	Hearing		Fever	Head ache	Head cold	Vomiting
			Unilateral	Bilateral	Male	Female	< 40 db	40—80 db	> 80 db	+	—	Unknown					Improved	Not improved				
Virus infection	Numb.	25	20	5	16	9	1	11	13	8	3	14	17	20	11	7	10	15	11	10	14	6
	Perc.		80	20	64	36	4	44	52	32	12	56	68	80	44	28	40	60	44	40	56	24
Probable virus infection	Numb.	23	17	6	13	10	1	10	12	7	2	14	16	15	13	10	8	15	17	7	16	6
	Perc.		74	26	57	43	4	44	52	30	9	61	70	65	57	43	35	65	74	30	70	26
Others	Numb.	62	54	8	40	22	4	30	28	14	17	31	45	38	28	16	29	33	1	7	6	2
	Perc.		87	13	65	35	7	48	45	23	27	50	73	61	45	26	47	53	2	11	10	3

must be present in the third group of table 8. That factors other than those mentioned by us can play a part in the etiology of sudden perceptive deafness can be neither denied nor affirmed.

Table 8 indicates that the number of patients for whom a virus infection can be established or presumed constituted 43 % of the total number of patients. The remaining patients undoubtedly, as stated already, include some undiscovered cases caused by virus. It can then be said that about half of the cases of acute perceptive deafness are probably due to a virus infection.

It is now important to investigate whether the symptomatology in the virus infection group differs from that of the remaining group. It would be natural to expect the infection connected with more binaural deafness and deafness with central localization, whereas in the case of circulatory disturbances, the expectation would be an unilateral involvement of the cochlea. It now appears that in the virus infections 77 % are localized unilaterally, and among the remaining cases 87 %. There is thus no well-defined difference.

Just as little difference is present according to data for the sexes, and the degree of deafness is approximately the same for both.

Fever was naturally more common in the group with confirmed and probable virus infections. The same applies to headache and cold symptoms. In the group of probable virus infections these symptoms were the criteria by which members were selected. The same applies to vomitting.

Dizziness combined with the deafness occurred in 50 % of the virus-infection group and in 45 % of the other. There is no clear difference here either.

Vestibulary lesions disclosed by the caloric test were present in 28 % of the virus group and 26 % of the others. In contrast, in the probable virus infection group not less than 43 % of the patients showed vestibulary lesions. The numbers are however too small to be conclusive.

The very important phenomenon of recruitment, which to some extent informs us whether the process is localized in the cochlea, was somewhat more frequent in the virus infection group.

Improvement in hearing was somewhat less frequent in the virus group than in the other group. This also applies to the diplacusis.

Summarizing, we can thus say that except for the meningeal irritation symptoms no important differences in symptomatology can be found between the virus group and the others.

X

PROGNOSIS

It is frequently assumed that the prognosis in this sudden perceptive deafness is very poor. It appears from this study, however, that such a somber view is not necessarily justified. The affliction is usually confined to one side, so that the ability to understand speech is retained. Out of our 110 patients, only 19 were affected on both sides. Two of these regained their hearing almost completely, and among the 17 remaining, 11 had a lasting hearing loss greater than 80 db and thus a nearly complete inability to understand speech.

The remaining 91 patients had a one-sided deafness which varied widely in degree. In this group there was a complete or almost complete recovery of 22 patients. For 36 patients a lasting loss of 40—80 db persisted, and for 33 there was a total loss of useful hearing. An improvement, to a greater or lesser extent, was therefore seen in 45 patients suffering from a unilateral deafness.

It can thus be said in general that the prognosis concerning recovery from the attacks of deafness is not bad.

A tendency toward spontaneous recovery of the hearing could be observed in 19 patients who were not treated. As reported also by other authors, this disease was never seen to take a fatal course.

An important question repeatedly asked by the patients was whether there was any danger that the other ear might now become affected. We closely questioned our 110 patients as to whether in the course of their lives they had at any time had a similar attack of deafness. This had been the case with only one patient, a woman of 52 years who had an acute perceptive deafness of both ears. In her 32nd year she had had a severe attack of grippe of the head and at that time became suddenly deaf with ringing in the right ear. She had heard a clap in her ear as she was blowing her nose and was thereafter deaf. The hearing had improved somewhat after 14 days. Fifteen years later, when we saw her, she had again had a grippe with high fever and could then suddenly hear nothing at all with either ear. This time the deafness did not go away.

As far as the other symptoms are concerned, 15 patients continued to a greater or lesser extent to complain of the tinnitus, while only one patient remained slightly dizzy.

XI

THERAPY

A adequate therapy for virus infections does not exist. We have only means with which to influence vasospasm and labyrinth hydrops. In most patients, however, it was not possible to determine the presence of these conditions with any degree of certainty. The effect of a therapy is difficult to ascertain because more than half of our patients showed spontaneous improvement, and frequently the loss of hearing varied in degree from day to day.

Among the cases of virus infections, 7 patients were treated with aureomycine and spasmolytics. During this medication, 4 patients did indeed show improvement, but never to such an extent that useful hearing was restored. Four patients who had "benign" meningitis and positive serological reactions were given 2 gr. Salycilas-Na. This improved the general condition of the patient, but a clear audiometric improvement was not achieved.

Spasmolytics (nicotinic acid and aminophylline) were given intravenously twice a day to 54 patients. If significant improvement could be shown afterwards, it cannot with any degree of certainty be ascribed to the therapy. Half of this group showed improvement, but seldom one of a lasting nature. The audiograms were taken directly before and ½ hour after the injection.

Anti-allergic and dehydration therapies were also attempted but without any results worth mentioning.

Supported by the literature and personal experience with the favorable effect of vitamin A in perceptive deafness, this drug was also used. The result however was not convincing in patients suffering from sudden perceptive deafness. It is particularly with such therapies involving extended periods of time that it is most difficult to determine to what extent improvement is spontaneous.

In 3 patients with a lowering of the heparine tolerance and reduction of the prothrombine time, dicumacyl was administered, however without effect.

In analogy with rheumatic facial paralysis for which cortisone is given to counteract the toxic oedema, it is certainly worth considering applying this therapy in the first days after the onset of the disease. Our experience is as yet insufficient to permit any judgement.

Just as all other investigators in this field, we continue to search for a therapy. We must consider that improvement, however scanty, can be of crucial importance to a deaf person who finds himself at the

borderline of useful hearing. A patient with a hearing loss of 70—80 db for whom a 10 db advantage can be obtained, will, with the help of a hearing aid, be in a far better position than before. When the loss is of 30—40 db, just such small gains are most important in whether or not a hearing aid is necessary.

TABLE 9

The effect of various medicaments on deafness.

Drug	Number	Improved	Unchanged	Worse
Aureomycine	4	—	4	—
Aureomycine + Spasmolytica	7	4	2	1
Euphylline	22	11	11	—
Ac. Nicotinicum	32	10	20	2
Dicumacyl	3	—	3	—
Salicyl natr.	4	—	4	—

XII

DISCUSSION

The main differences in opinion between the investigators of sudden perceptive deafness concern two questions, its etiology and the localisation of the lesions.

As a result of study of the literature and our own experience, we are certain that several different causes of this illness must be assumed. One of these causes is virus infection, and especially reinfection with mumps. The only author who mentions this possibility is COOMBS, and we are glad that we can confirm his opinion. As to the importance of virus infections, there are good reasons to assume that more than half of our cases are caused by such an infection.

For the remaining cases, we are just as uncertain as all the other investigators. We too supposed vascular disturbances or abnormal coagulation and also endolymphatic hydrops or unknown intoxications to play a role. As compared to the clearly demonstrated virus infections, all these suppositions are highly unsatisfactory. This is all the more cogent because we could gather arguments in favour of the unimportance of vasospasm and blood coagulation abnormalities, except in the case of angiosclerosis in older people. Nor can enough evidence for these other suppositions be found in literature. For instance, FOWLER ascribed the highest importance to studying the blood sludging caused by emotional factors, but he could demonstrate this phenomenon in only one patient. MYGIND, HALLBERG and others lay much stress on labyrinthine hydrops as in Ménière's desease, although fenestration operations and dehydration failed to prove this theory.

In accordance with those who think that spasm of the end-arteries of the cochlea could cause the deafness, we too administered vasodilatating drugs. We did indeed see some effects, but they were not lasting. Thus it is quite possible not that a spasm was removed but that a better circulation in the cochlea improved an oedema or a toxic condition.

As to the localisation of the lesions, a sharp distinction must be made between the virus infection and the other causes. In virus infections the localisation of the lesions must be based on what we know from obductions in mumps, poliomyelitis, etc. From this we may conclude that the lesions can be peripherical as well as central. This assumption is in accordance with the results of our audiometrical and vestibular findings.

As to the localisation of the process in the remaining cases, we are much in doubt. The expectation that we would find mainly peripherical

lesions was not confirmed. In fact we found the same conditions as in the virus infections cases: probable peripherial as well as central lesions. These facts could be best explained if general toxic agents acting on the nerve cells and the blood vessels are accepted as causes. Such a theory closely approaches the assumption of a circulating unknown virus causing damage in regions that have a special predisposition, as suggested by Coassolo for the rheumatic facial paralysis.

For this reason, we must hope that when new viruses are discovered more cases of sudden deafness can be classified among this group and other research will establish the cause in the remaining unknown cases.

XIII

SUMMARY

While observing two patients in February, 1954, who were suffering from sudden perceptive deafness together with meningitic irritation symptoms and from whose faeces and cerebro-spinal fluid the Columbia S.K. virus was isolated, we came to the opinion that virus infections in general might be an important etiological factor in this form of deafness. For this reason, such a virus infection was systematically sought for in these cases.

It is known that intoxication and some infectious diseases can result in such sudden loss of hearing. Sudden fluctuations in hearing for example are common in Ménière's desease. We have not included this category of deafness, in which the cause is known, in our study. This is contrary to other authors such as HALLBERG.

Of the 110 patients collected from 1954 through AUGUST, 1957, 74 were observed in our clinic and 36 obtained by means of an inquiry conducted among Dutch Ear, Nose and Throat specialists. The total group included 68 men and 42 women whose average age was 42 years.

Among these cases were only 19 with bilateral perceptive deafness, of whom 11 had a hearing loss on both sides of more than 80 db, so that they lacked all serviceable hearing. The remaining 8 were to a lesser degree deaf for speech. In the 91 unilateral cases, the understanding was not disturbed since hearing in the other ear was still adequate. The degree of deafness in the affected ear varied from 30 db to total loss. The recruitment symptom which was investigated in 51 patients was present in 29. Improvement in the hearing function itself was demonstrated in 45 patients, 2 of whom had a bilateral deafness, and spontaneous improvement was shown in 19 untreated cases.

The very annoying symptom of tinnitus, by which the patient's attention is sometimes drawn to his deafness, was present in 78 patients. It preceded deafness in 17 cases. In almost half of these patients the tinnitus disappeared within 1 month and it persisted in only a few of the remaining cases. Like tinnitus, as a symptom of cochlear lesion, diplacusis was reported by 73 patients when questioned concerning their symptoms. In 4 this symptom had led to awareness of their deafness. This symptom is also temporary, and was only reported by 2 patients as seemingly permanent.

Dizziness appeared in the histories of 52 of our patients. A caloric investigation of the labyrinth was done on 71 patients, of whom 10 were bilaterally deaf. The results of this investigation showed that 33 patients

exhibited a functional disturbance of the labyrinth, varying between slight deviation and total disability, while a nystagmus prepondorance was frequently evident. Spontaneous nystagmus was seen in only 19 patients suffering from unilateral deafness. It was noteworthy that 3 patients, in whom a sub-total deafness was measured in both ears, normally-reacting labyrinths were found and that in 5 only one labyrinth was affected.

In those cases showing fever and meningitic irritation together with a grippe-like infection, our conception that a virus infection might be the cause was indeed confirmed. Headache and nasal catarrh were often early symptoms in these cases. Abnormalities in the cerebro-spinal fluid, indicating a "benign" serous meningitis, was found in 14 of these patients. In 74 patients in whom the onset was reasonably recent, virological investigation was carried out. Virus was grown from the faeces and spinal fluid in two patients. In 25 cases positive serological reactions were found and among them, in not less than 14 cases, for the mumps virus. Only one patient had a mild parotitis epidemica. All of them had suffered from mumps in their childhood. Thus in these cases a reinfection must be accepted. Actual contact with individuals suffering from mumps was known for only 6 patients. The occurence of mumps reactions in a third of the cases of virus infections is statistically significant. Thus WOLFF has shown that among normal subjects positive mumps reactions are found in only $\pm$ 0.1 %, while individuals in contact with mumps cases showed 99.3 % positive reactions.

In the 7 positive reactions found for the "Enteric group", that is, 5 for Columbia S.K. and 2 for the Coxsackie group, the causal connection is not so definite but is very probable.

For the influenza group there were 7 positive reactions found, but this figure is certainly not significant since a similar number can be expected among normal subjects.

Previous investigators have not sufficiently distinguished between the etiology and pathogenesis of acute perceptive deafness. As a result, circulatory disturbances and labyrinthine hydrops have often been stipulated as causes, while it must be assumed that a deeper cause must underly these pathogenetically important mechanisms. Thus oedema and abnormalities in the blood vessels will always be found together with infection. In only two of our patients, who had cardio-vascular symptoms, did we suspected a vascular cause such as spasm or thrombosis. In approximately half of our 110 patients the etiology remained unknown. It is extremely probably that among these patients unknown or undetectable virus infections are an important factor.

For the localization of the process of this disease, a cochlear lesion was accepted in those cases in which recruitment, diplacusis and symptoms of a periferal vestibulary lesion were present. This does not exclude the possibility that, especially in the cases of virus infection,

a combination with a neuritis and encephalitis is very probable.

Therapy in the sudden perceptive deafness caused by virus infections is ineffective. Aureomycine and salicylas natricus were tried without result. On the supposition that in many cases, and even in infection, vasospasm plays a role, spasmolytics such as nicotinic acid and aminophylline were administered. Audiometrically checked immediate improvements were repeatedly observed. These improvements were, however, seldom lasting, and when lasting the possibility of a spontaneous recovery must be kept in mind.

The prognosis of this sudden perceptive deafness is, as far as the social hearing ability is concerned, not so poor as has been thought up till now. Of our 110 patients, 91 suffered from unilateral deafness. Among these, 53 were totally deaf at the onset but 22 recovered. The remaining 31 could hear sufficiently with their normal ear. Of the 19 patients suffering from bilateral deafness, 2 recovered completely and 6 needed a hearing aid. However, 11 remained totally deaf. This is 10 % of the total group of our patients.

It is estimated that in The Netherlands each year 200 cases of sudden perceptive deafness occur in a population of eleven million. It is of course probable that epidemics would increase this number.

RÉSUMÉ

A la suite de l'observation en Février 1954 de deux malades atteints de surdité de perception subite accompagnée de symptomes de réaction meningée et chez lesquels l'examen des matières et du liquide céphalo-rachidien a révélé la présence du virus Columbia S.K., nous avons été amenés à penser que les infections virusales en général pourraient être un facteur étiologique important dans cette forme de surdité. Nous avons donc recherché systématiquement une telle infection virusale.

On sait que des intoxications et certaines maladies infectieuses peuvent provoquer une telle perte subite de l'ouie. Des variations subites de l'ouïe apparaissent également régulièrement dans la maladie de Ménière. Contrairement à d'autres auteurs, tels que HALLBERG, nous n'avons pas inclus dans notre étude ces cas dont l'étiologie est donc connue.

Sur les 110 malades rassemblés de 1954 à Août 1957 compris, 74 ont été observés cliniquement par nous et 36 nous ont été fournis à la suite d'une enquête auprès des oto-rhino-laryngologistes néerlandais. Le groupe comprenait au total 68 hommes et 42 femmes dont l'âge moyen était de 42 ans.

Parmi eux ne se trouvaient que 19 malades atteints de surdité de perception bilatérale, dont 11 chez qui la perte de l'ouïe des deux côtés dépassait 80 dB, ce qui signifie une surdité à peu près totale.

Les 8 autres malades étaient dans une mesure moindre sourds à la parole. Dans les 91 cas de surdité unilatérale, la compréhension du

langage parlé n'était pas altérée, l'acuité auditive étant toujours restée suffisante dans l'autre oreille. Le degré de surdité de l'oreille atteinte variait d'une perte de 30 dB à la perte totale, de l'ouïe. Le symptome de recruitment recherché chez 51 malades a été trouvé présent chez 29 d'entre eux. En ce qui concerne la fonction auditive, on a pu déceler une amélioration chez 45 malades, dont 2 atteints de surdité bilatérale et l'on a pu constater une tendance à l'amélioration spontanée chez 19 d'entre eux.

Les tintements très désagréables par lesquels se signale quelquefois la surdité étaient présents chez 78 malades. La surdité avait été précédée de ce symptome chez 17 d'entre eux et remarquée plus tard seulement par 5 d'entre eux. Chez la moitié environ de ces malades, les tintements avaient disparu au bout d'un mois et n'avaient persisté que chez quelquesuns des autres malades.

En même temps que les tintements, un autre symptome de lésion cochléaire, une diplacusie avait été signalée par 73 malades au cours de l'interrogatoire. Chez 4 malades, c'est ce symptome qui leur avait fait remarquer leur surdité. Ce symptome est lui aussi transitoire et n'a été signalé comme permanent que par 3 malades.

Des vertiges étaient présents dans l'histoire de la maladie de 52 de nos malades. L'épreuve calorique du labyrinthe a été pratiquée chez 71 malades dont 10 cas de surdité bilatérale. Cette épreuve a montré qu'il existait chez 33 malades un dérèglement de la fonction du labyrinthe allant d'une atteinte légère à un arrêt total, tandis que l'on a pu déceler souvent la présence d'une tendance au nystagmus. Un nystagmus spontané n'a été constaté que chez 9 malades atteints de surdité unilatérale. Assez curieusement, 3 malades révélant à l'examen une surdité subtotale des deux oreilles avaient des labyrinthes réagissant normalement et 5 autres ne présentaient qu'une atteinte labyrinthaire unilatérale.

Dans les cas accompagnés de fièvre, de réaction méningée et d'une infection de type grippal, notre supposition qu'une infection virusale était à l'origine de la surdité s'est trouvée effectivement confirmée. Les premiers symptomes avaient souvent été chez ces malades une céphalée et un catarrhe nasal. Des anomalies du liquide céphalo-rachidien faisant penser à une méningite séreuse bénigne ont été constatées chez 14 de ces malades. Chez 74 malades dont le début de la maladie n'était pas ancien, il a été procédé à un examen virologique. Chez deux d'entre eux, on a pu cultiver un virus dans les matières et le liquide céphalo-rachidien. Dans 25 cas, les réactions sérologiques ont été positives, dont 14 fois pour le virus des oreillons.

Un malade seulement présentait une atteinte légère de cette maladie. Tous avaient déjà eu les oreillons, d'où il s'ensuit que nous avons ici à faire à une réinfection. Six malades seulement savaient avoir été en contact avec des oreilloneux.

La présence d'une réaction positive au virus des oreillons dans un

tiers des cas d'infection virusale est statistiquement significative pour la valeur d'agent pathogène du virus des oreillons. En effet WOLFF a montré que chez les individus normaux, la réaction du virus des oreillons n'est positivé que dans 0,1 % des cas, tandis que chez des personnes ayant été en contact avec des oreilloneux on a pu constater 99, 3 % de réactions positives.

Pour les 7 réactions positives pour l'„enteral group" constatées (5 pour Columbia S.K. et 2 pour le groupe Coxsackie), la relation causale est vraisemblable. Pour le groupe de l'influenza, on a constaté 7 réactions positives mais ce chiffre n'est pas significatif car on trouve chez les individus normaux un pourcentage à peu près équivalent. L'étiologie et la pathogénèse de la surdité de perception aigüe n'ont souvent pas été suffisamment distinguées jusqu'ici. Ainsi on en avait souvent recherché l'origine dans des troubles circulatoires et une hydropsie labyrinthaire, alors qu'il faut bien admettre qu'une cause plus profonde est à la base de ces mécanismes si importants pour la pathogénèse. Car on trouve toujours en cas d'infection des oedèmes et des atteintes vasculaires. Chez 12 de nos malades seulement qui présentaient des symptomes cardio-vasculaires, une origine vasculaire, telle que spasme ou thrombose a été envisagée. Pour la moitié environ de nos 110 malades l'étiologie est restée inconnue. Il est fort possible que parmi ces malades, des infections virusales inconnues ou non reconnues aient pu être un facteur important.

Quant à la localisation du processus de cette maladie, les nombreux cas où apparaissaient recruitment, diplacusie et symptomes de lésion vestibulaire périphérique amènent à admettre une lésion cochléaire. Ceci n'exclut pas, surtout dans les cas d'infection virusale, qu'une combinaison avec une névrite et une encéphalite soit très vraisemblable.

La thérapeutique de la surdité de perception brusque causée par des infections virusales est impuissante. L'auréomycine, le salicylate de soude ont été essayés sans résultats appréciables. Comme on avait supposé que dans de nombreux cas et même en cas d'infection virusale, des spasmes vasculaires jouaient un rôle, on a administré des spasmolytiques tels que l'acide nicotinique et l'aminophylline. Des améliorations directes contrôlées, audiométriquement ont été constateés à diverses reprises. Cependant ces améliorations n'étaient généralement pas durables, et lorsqu'elles l'étaient, il convient de tenir compte de la possibilité de rémissions spontanées.

Le pronostic de ces surdités de perception subites n'est, en ce qui concerne la validité sociale, pas ausi mauvais qu'on le pensait jusqu'à présent. Sur nos 110 malades, 91 étaient atteints de surdité unilatérale. De ces malades, 53 étaient dès le début de leur maladie totalement sourds; mais 22 s'étaient remis. Les 31 malades restants entendaient bien avec leur oreille normale. Sur les 19 malades atteints de surdité bilatérale, 2 s'étaient complètement remis et 6 entendaient avec l'aide

d'une prothèse auditive. Onze malades sont restés totalement sourds, ce qui représente 10 % du nombre total de nos malades.

En Hollande il y a annuellement environ 200 cas de surdité de perception subite sur une population de 11 millions d'habitants. Naturellement il est possible qu'en cas d'épidémies ce nombre augmente temporairement.

ZUSAMMENFASSUNG

Durch Beobachtung von zwei an plötzlicher Perzeptionstaubheit, zusammen mit meningealen Reizerscheinungen leidenden Patienten im Februar 1954, bei denen aus den Fäkalien und dem Liquor der Columbia S.K. Virus isoliert wurde, kam bei uns der Gedanke auf, dasz Virusinfektion im Allgemeinen bei diezer Art der Taubheit eine bedeutende ätiologische Erscheinung sein könnte. Deshalb wurde systematisch nach einer Virusinfektion gesucht.

Es ist bekannt, dasz Intoxikationen und verschiedene Infektionskrankheiten einen solchen plötzlichen Gehörverlust zur Folge haben können. Auch kommen solche plötzlichen Schwankungen im Gehör bei der Ménièrischen Krankheit regelmäszig vor. Diese Fälle, deren Ursache demnach bekannt ist, haben wir im Gegensatz zu anderen Autoren, wie HALLBERG, nicht in unsere Untersuchung mit einbezogen.

Von den seit 1954 bis August 1957 erfassten 110 Patienten wurden von uns 74 klinisch observiert und 36 durch eine Umfrage bei niederländischen Hals-, Nasen- und Ohrärtzten erhalten. Die Gruppe umfaszte 68 Männer und 42 Frauen im Durchschnittsalter von 42 Jahren.

Unter diesen befanden sich nur 19 an bilaterale Perzeptionstaubheit leidende Personen, davon 11 mit einem Gehörverlust an beiderseits gröszer als 80 dB, d.h. nahezu völliger Taubheit. Die übrigen 8 Patienten waren in geringeren Masze Sprachtaub. Bei den 91 Fällen unilateraler Taubheit war das Sprachgehör nicht gestört, weil im anderen Ohr die Hörschärfe stets genügend geblieben war. Der Taubheitsgrad des kranken Ohres variierte von 30 dB Verlust bis zum gänzlichen Ausfall des Gehörs.

Von 51 Patienten zeigten 29 das Lautheitsausgleichsymptom. Bei 45 Patienten, darunter zwei mit einer bilateralen Taubheit, wurde eine Verbesserung der Gehörsfunktion festgestellt; bei 19 von ihen hatte sich bereits spontan eine Besserung gezeigt.

Die sehr lästige Tinnituserscheinung, wodurch manchmal die Taubheit bemerkt wird, war bei 78 Patienten vorhanden. Bei 17 von ihen ging dieses Symptom der Taubheit voraus und nur bei 5 wurde es erst später bemerkt. Der Tinnitus verschwand bei ungefähr der Hälfte der Patienten innerhalb eines Monats und nur in einigen der übrigen Fällen blieb dieser bestehen.

Ebenso wie Tinnitus wurde als Symptom einer cochleären Laesion von 73 Patienten bei Nachfrage Diplacusis in der Anamnese angegeben.

Bei 4 Patienten wurde auf Grund dieser Erscheinung die Taubheit bemerkt. Auch dieses Symptom tritt vorübergehend auf und wurde nur von 2 Patienten als bleibend bezeichnet.

In der Krankheitsgeschichte kamen bei 52 unserer Patienten Schwindelerscheinungen vor. Die kalorische Untersuchung des Labyrinths wurde bei 71 Patienten, worunter 10 doppelseitigen Taubheitsfällen, vorgenommen. Die Untersuchung ergab bei 33 Patienten eine Funktionsstörung des Labyrinths, varierend von einer leichten Abweichung bis zum völligen Ausfall, während oft eine Nystagmusbereitschaft festgestellt werden konnte. Ein spontaner Nystagmus wurde nur bei 9 an einseitigen Taubheit leidende Patienten gesehen. Merkwürdig was, dasz bei 3 Patienten, bei denen eine subtotale Taubheit beiden Ohren fastgestellt worden war die Vestibulärorgane normal reagierten und bei 5 anderen das Labyrinth einseitig gestört war.

In den Fällen mit Fieber und meningealen Reizerscheinungen, wurde unsere Vermutung einer Virusinfektion bestätigt. Kopfschmerzen und Schnupfen waren dabei oft die ersten Symptome. So wurden Veränderungen des Liquor cerebrospinalis, die auf eine gutartige seröse Meningitis wiesen, bei 14 dieser Patienten gefunden. 74 (worunter 48 virus-verdächtige) Patienten, bei denen die Erkrankung nicht lange zurück lag, wurden virologisch untersucht. Aus den Fäkalien und dem Liquor konnte nur bei zweien Virus isoliert werden. In 25 Fällen waren die serologischen Reaktionen positiv und zwar in nicht weniger als 14 Fällen für das Mumpsvirus. Nur 1 Patient litt an einer leichten Parotitis epidemica. Alle hatten bereits früher Mumps gehabt, sodasz es sich hier um eine erneute Infektion handelte. Ein Kontakt mit Mumpskranken war nur 6 Patienten bekannt. Dieses Auftreten der Mumpsreaktionen in einem Drittel der Fälle von vermütlichen Virusinfektionen ist statistisch gesehen, beweisend für die ursächliche Bedeutung des Mumpsvirus. Durch WOLFF ist ja bewiesen worden, dasz bei normalen Versuchspersonen positive Mumpsreaktionen nur bei $\pm$ 0,1 % vorkommen. Dahingegen zeigten sich bei Personen, die mit Mumpspatienten in Berührung gewesen waren, bei 99,3 % positive Reaktionen.

Bei den 7 für die "Enterale Gruppe" festgestellten positiven Reaktionen, nämlich 5 für Columbia S.K. und 2 für die Coxsackie-Gruppe ist der ursächliche Zusammenhang wahrscheinlich.

In der Influenza-Gruppe wurden 7 positive Reaktionen gefunden, jedoch ist diese Anzahl sicher nicht bezeichnend, da unter normalen Versuchspersonen ebenfalls eine ungefähr gleiche Anzahl zu erwarten wäre.

Durch andere Forscher ist die Ätiologie und Pathogenese der akuten Perzeptionstaubheit oft nicht genügend erkannt worden. So wurden als Ursache oft Kreislaufstörungen und labyrinthärer Hydrops angenommen, während doch angenommen werden musz, dasz diesen pathogenetisch wichtigen Mechanismen eine tiefere Ursache zugrundeliegt.

So wird man bei Infektionen stets Oedem und Gefäszveränderungen finden. Nur bei 12 unserer Patienten mit cardiovasculären Symptomen wurde an eine vasculäre Ursache, wie Spasmus und Thrombose gedacht. Es ist sehr wohl möglich, dasz bei vielen Patienten unbekannte oder nicht erkannte Virusinfektionen eine wesentliche Rolle spielen.

Der Krankheitsprozess musz aufgrund vieler Fälle, in denen Lautheitsausgleich, Diplacusis und Symptome einer periferen vestibulären Läsion vorhanden waren, cohleär lokalisiert werden.

Dies schlieszt jedoch nicht aus, dasz, vor allem in Fällen von Virusinfektion, ein Zusammenhang mit einer Neuritis und Encephalitis sehr wahrscheinlich ist.

Die Therapie der durch Virusinfektion verursachten plötzlichen Perzeptionstaubheit ist wirkungslos. Aureomycin und Natriumsalicylat wurden ohne deutlichen Erfolg verabreicht. In der Annahme, dasz in manchen Fällen und sogar auch bei Infektion Gefäszspasmen eine Rolle spielen, wurden Spasmolytica, wie Nikotinsäure und Aminophyllin versucht. Wierholt konnten audiometrisch kontrollierte direkte Besserungen wahrgenommen werden. Diese Besserungen waren jedoch meistens nicht von bleibender Art, und wenn letzteres der Fall war, musz mit der Möglichkeit der spontanen Remission gerechnet werden.

Die Prognose dieser plötzlichen Perzeptionstaubheit ist im Hinblick auf die soziale Validität nicht so ungünstig wie bisher angenommen. Von unseren 110 Patienten litten 91 an einer unilateralen Taubheit. Darunter waren 53 zu Beginn vollständig taub. 22 erholten sich gänzlich. Die übrigen 31 konnten mit dem gesunden Ohr gut hören. Von den 19 bilateral tauben Patienten genasen 2 völlig; das Gehör 6 anderer konnte mit Hilfe eines Hörapparates verbessert werden. 11 blieben jedoch vollständig taub. Dies bedeutet also 10 % aller unserer Patienten.

Schätzungsweise kommt in den Niederlanden auf eine Bevölkerung von elf Millionen jährlich eine Anzahl von 200 dieser Fälle plötzlicher Perzeptionstaubheit vor. Es ist möglich, dasz bei Virus-epidemien diese Anzahl eine vorübergehende Zunahme erfahren wird.

RESUMEN

Come resultado de las observaciones llevadas a cabo en Febrero de 1954 en dos pacientes que sufrían de sordera perceptiva súbita, conjuntamente con numerosos síntomas de prurito, y en los cuales se aisló el virus Columbia S.K. en las heces fecales y las secreciones, llegamos a pensar, que en general, el virus infeccioso en esta forma de sordera bien pudiera ser un factor etiológico importante. Esto condujo pues a iniciar sistemáticamente la búsqueda de ese virus infeccioso.

Es sabido, que en algunas enfermedades infecciosas las intoxicaciones pueden provocar súbitamente una sordera semejante. Igualmente ocurren con regularidad, en la enfermedad de Ménière, las oscilaciones

súbitas en la audición. A diferencia pues de otros autores, entre ellos HALLBERG, no hemos incluído en nuestras investigaciones estos casos, en los cuales la causa es ya conocida.

De los 110 pacientes — catalogados desde 1954 hasta el mes de Agosto de 1957 incluído — observamos clínicamente a 74 de ellos y el historial clínico de los restantes fué obtenido mediante una encuesta entre los médicos otorinolaringólogos holandeses. Constituían este grupo 68 hombres y 42 mujeres, con un promedio de edad de 42 años.

Entre ellos se encontraban solamente 19 pacientes padeciendo de una sordera perceptiva bilateral, 11 de éstos con la pérdida de la audición en ambos oídos en un grado de 80 dB, es decir, punto menos que completamente sordos. Los 8 pacientes restantes eran ligeramente sordos a la palabra hablada (escuchar). Entre las 91 sorderas unilaterales, no estaba perturbada o interrumpida la facultad de escuchar, puesto que en el otro oído la percepción auditiva era suficiente aún. El grado de sordera en el oído afectado variaba entre una pérdida de 30 dB hasta un ensordecimiento total. El síntoma de reclutamiento, estudiado en 51 pacientes, se hallaba presente en 29 de estos casos. En lo que se refiere a la función auditiva, pudo comprobarse una mejoría en 45 pacientes, entre ellos 2 también con una sordera bilateral y en otros 19, una inclinación espontánea de mejoría.

El fenómeno de tinnitus, tan molesto, y mediante el cual puede a veces comprobarse la sordera, se hallaba presente en 19 pacientes. En 17 de ellos este síntoma precedió a la sordera y sólo en otros 5 fué observado más adelante. En la mitad aproximada de estos pacientes, el tinnitus desapareció en el término de un mes y apenas en alguno de los restantes persistió este fenómeno.

A semejanza que el tinnitus, como síntoma de una lesión coclear, la diploacusia apareció en la historia clínica de 73 pacientes, cuando fueron interrogados una vez más. En 4 pacientes fué este mismo síntoma la causa de que observaran su sordera. Este síntoma es transitorio también y sólo fué citado como permanente por 2 pacientes.

Los vértigos aparecieron en la historia clínica de 52 de nuestros pacientes. El reconocimiento calórico del laberinto se llevó a cabo en 71 pacientes, entre los que se hallaban 10 sufriendo de sordera bilateral. El resultado de este reconocimiento demostró en 33 pacientes una perturbación en la función del laberinto, que variaba desde una ligera irregularidad hasta una supresión completa, en tanto que con frecuencia pudo evidenciarse un nystagmus de preferencia. Un nystagmus espontáneo se observó escasamente en 9 pacientes sufriendo de una sordera unilateral. Es singular, que en 3 pacientes en los que pudo medirse una sordera semitotal en ambos oídos, se comprobara la reacción normal de los laberintos, y en otros 5 sólo estaba afectado un laberinto.

En los casos en que la fiebre y síntomas de prurito meníngeos, y una infección gripal se hallaban presentes, quedó confirmada nuestra

sospecha de la presencia de un virus infeccioso. Dolores de cabeza y catarros nasales se presentaron con frecuencia como síntomas preliminares. Así se descubrieron en 14 de estos pacientes, irregularidades en el líquido cerebroespinal, que indicaban una meningitis serosa benigna. Se hizo una búsqueda de virus en 74 pacientes, en los cuales el proceso estaba en su comienzo. De las heces fecales y el liqor de 2 de ellos, se cultivó el virus. En 25 casos las reacciones serológicas fueron positivas y en no menos de 14 casos acusaron el virus parotídes. Sólo 1 paciente padecía una parotiditis epidémica. Todos estos pacientes habían pasado ya esta enfermedad y se trataba en estos casos de una nueva infección. De 6 pacientes únicamente se sabía que habían estado en contacto con otros pacientes sufriendo de parotiditis. La presencia de esta reacción parótida, en un tercio de las infecciones virulentas, es significativa — estadísticamente considerada — para la significación causativa del virus parotídes. WOLFF ha probado además, que entre personas normales que voluntariamente se han prestado a las observaciones, las reacciones positivas de virus de la parotiditis se presentan escasamente en un 0.1 % aproximadamente. Mas, entre personas que estuvieron en contacto con pacientes sufriendo de parotiditis, las reacciones positivas alcanzaron un procentaje de un 99.3 %.

En las 7 reacciones positivas, demostradas para el "Grupo Enteral", es decir, 5 para Columbia S.K. y 2 para el grupo Coxsackie, la relación causal es aparente.

Para el grupo de influenza se encontraron 7 reacciones positivas. En realidad este número carece ciertamente de significación, puesto que en las pruebas entre personas normales, puede esperarse así mismo un parecido número de iguales reacciones.

Investigadores que realizaron previamente estos estudios, no pudieron diferenciar con frecuencia suficientemente la etiología y patogénesis de a sordera perceptiva aguda. Así se ha aceptado con frecuencia comol causa de ello, los trastornos de la circulación y las secreciones del laberinto, en tanto que debemos reconocer no obstante, que en este importantísimo mecanismo patógeno se halla una razón aún más fundamental, puesto que en cialquier infeccion podemos comprobar la presencia de edemas y trastornos vasculares. Sólo en 12 de nuestros pacientes, que presentaban síntomas cardiovasculares, se pensó en una causa vascular, tal como el espasmo y la trombosis. En la mitad aproximada de nuestros 110 pacientes, la etiología permaneció desconocida. Es muy posible que entre estos pacientes, infecciones virulentas desconocidas o no reconocidas aún, puedan constituir un factor importante.

Con respecto a la localización del proceso en esta enfermedad, creemos que las lesiones cocleares juegan un papel de importancia debido a la existencia de numerosos casos que presentaban síntomas de lesión en la periferia vestibular asi como diploacusia y reclutamiento. Esto no excluye ciertamente, en especial en los casos de infecciones

virulentas, la posibilidad de una neuritis y encefalitis combinada.

La terapia en una sordera perceptiva súbita, es inútil, cuando esta sordera es provocada por infecciones virulentas. Aureomicina y salicylas natricus, han sido empleadas sin resultado alguno efectivo. En vista de que se supone que en muchos casos y también en las infecciones los espasmos vasculares desempeñan su parte, se han administrado espasmódicos tales como ácido nicotínico y aminofilina. La comprobación audiométrica acusó repetidas mejorías directas. Estas mejorías sin embargo no poseían un carácter permanente y dado el caso de que resultaran permanentes, siempre había que tener en cuenta la posibilidad de recaídas espontáneas.

El pronóstico de estas sorderas perceptivas súbitas no es, en lo que respecta al valor social del individuo, tan malo como hasta ahora se suponia. 91 de nuestros 110 pacientes padecen una sordera unilateral. 53 de ellos, estaban totalmente sordos desde el comienzo, de los caules 23 se recuferaron. Los 31 restantes pueden oir bien con el oido normal. De los 19 pacientes que sufrían una sordera bilateral, 2 se recuperaron completamente y 6 mejoraron con ayuda de un aparato. Sin embargo, quedan 11 completamente sordos. Esto representa solamente el 10 % de nuestros pacientes.

Según los cómputos verificados, cada año, existen en Holanda unos 200 casos de esta sordera perceptiva súbita, en una población de once millones de habitantes. Es lógico que durante las epidemias este número aumente durante algún tiempo.

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STELLINGEN

I

Meer dan de helft van de gevallen van plotselinge perceptie-doofheden wordt veroorzaakt door virusinfecties en wel meestal door reinfectie met het bofvirus.

II

Voor de pathogenese van de plotselinge perceptie-doofheid moet aangenomen worden toxische beschadiging van de cochlea zowel als encephalitische processen door goedaardige meningitis.

III

De acute perceptie-doofheid dient onderscheiden te worden van de ziekte van Ménière. Bij de eerste speelt endolymphatische hydrops vermoedelijk geen of een zeer geringe rol.

IV

Er zijn geen aanwijzingen dat patiënten lijdende aan allergie en vaatspasmen gepraedisponeerd zijn voor het verkrijgen van een plotselinge perceptie-doofheid.

V

De prognose van de plotselinge perceptie-doofheid is in vergelijk met andere perceptie doofheden gunstig, ook wat betreft tinnitus en duizeligheid. Herhaling van de aanval is uiterst zeldzaam.

VI

Het gebruik van het woord nystagmus voor andere delen van het lichaam dan het oog, verdient geen aanbeveling.

VII

Bij hoofdpijn als gevolg van een lumbaalpunctie geve men beter vruchtensappen dan water.

VIII

Het gebruik van een dikke naald bij lumbaalpunctie is af te keuren.

IX

Alhoewel een aantal patiënten, lijdende aan de ziekte van Ménière, goed reageren op antihistaminica, moet een allergische oorzaak in de meeste gevallen onwaarschijnlijk geacht worden.

X

Voor locale antibiotische behandeling op huid, conjunctivae en in oren neme men die antibiotica die voor interne toepassing niet gebruikelijk zijn.

XI

Bij het streven tot het op peil brengen van het onderwijs in Suriname, is een uitwisseling van leerkrachten een dringende noodzaak.

XII

Bij de behandeling van het carcinoom van de bovenkaak (sinus maxillaris) verdient het aanbeveling de bestraling met snelle electronen in overweging te nemen.

XIII

In verband met de uitgesproken domestische levenswijze en het beperkte vliegbereik van de adulte Culex quinquefasciatus, kan worden aangenomen dat de sterk uiteenlopende percentages microfilariaemie onder de Creolenbevolking in Suriname voornamelijk het gevolg zijn van verschillen in de bevolkingsdichtheid.