THE CLINICAL SIGNIFICANCE OF ELECTROCOCHLEOGRAPHY

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SIETZE JOHANNES RIETEMA

The clinical significance of electrococleography

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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. D.J. KUENEN, HOOGLERAAR IN DE FACULTEIT DER WISKUNDE EN NATUURWETENSCHAPPEN, VOLGENS HET BESLUIT VAN HET COLLEGE VAN DEKANEN TE VERDEDIGEN OP DINSDAG 5 JUNI 1979 TE KLOKKE 15.15 UUR.

Door

SIETZE JOHANNES RIETEMA
geboren te Stadskanaal in 1946

Promotoren: DR. J.J. EGGERMONT
PROF. DR. P.H. SCHMIDT

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Aan Hanke

Contents

Abbreviations and definitions

ECoG : electrocochleography
AP : action potential
CM : cochlear microphonics
SP : summating potential

peripheral hearing loss is caused by a lesion located outside the pia mater; the lesion can be located in

a) the external ear
b) the middle ear } conductive hearing loss

c) the cochlea
d) the retrocochlear area } perceptive hearing loss

central hearing loss is caused by a lesion located within the pia mater

1 Introduction

Almost two hundred years after Galvani's discovery of the electrical characteristics of biological tissue, the electrocardiogram and the electroencephalogram are well established methods of clinical examination. In 1930, Wever and Bray discovered the electrical activity in the inner ear, which was called the cochlear microphonics. However, the development of an electrical method for investigation of the functioning of the human inner ear has taken a much slower course than that for the electrocardiogram and the electroencephalogram.

The first round window recording of the cochlear microphonics (CM) in man was performed five years after Wevers' discovery of the phenomenon (Fromm et al, 1935). Although the recording techniques improved, so that better CM recordings could be produced, it lasted until the early fifties before recordings of the compound action potential (AP) could be reliably obtained in animal experiments (Tasaki et al, 1954). After 1960, a divergence took place: some investigators concentrated on single fiber recordings in animals (Kiang et al, 1965), while others gave their attention and energy to AP measurement in man (Ruben et al, 1960, 1962). These AP recordings from the round window were all made during surgery; improvement of the recording techniques led to clear click-evoked compound action potentials at high stimulus intensities. Ruben (1967) indicated three important topics of electrocochleography (ECoG):

1. the correlation of physiological and psychoacoustic properties,
2. the investigation of certain diseases, and
3. the objective diagnosis of individual cases of deafness.

The last two topics especially are important in the clinical use of ECoG.

In 1967, when measurements in man equalled the level of sophistication of the animal experiments in the early fifties, electrocochleography became a routine diagnostic procedure. This step was taken independently by Japanese (Yoshie et al, 1967) and French (Portmann et al, 1967) scientists. Yoshie recorded from the outer ear canal, but later also used the promontory recording. The use of average response computers for routine in electrocochleography has made it possible to record compound action potentials in response to stimuli having an intensity close to the subjective hearing threshold.

Both the use of the average and of the transtympanic or extratympanic approach have converted the method from an operation-room procedure to a routine office procedure. (Montandon et al, 1975).

With the use of tone-bursts (Yoshie, 1973, Eggermont et al, 1974) as stimulus along with the well known click, and the introduction of more sophisticated techniques (e.g. high-pass noise masking (Elberling, 1974; Eggermont et al. 1976)) clinical elec-

trocochleography reached beyond the level at which the animal experiments had halted.
Because the characteristics of single fiber responses became available (Kiang et al, 1965) together with information about the anatomical substrate of cochlear microphonics and summating potential responses (Whitfield and Ross, 1965, Dallos et al, 1972 b) electrocochleographic results could now be interpreted more accurately than the animal experimental results in the late fifties.
The application of electrical responses in clinical audiology had started with the recording of the 'slow' vertex potentials. The clinical usefulness of this method has been limited, especially in children who cannot be sedated adequately without influencing the responses (Davis, 1976, Schmidt and Spoor, 1974).
The responses recorded in electrocochleography are on the other hand peripheral responses which are not influenced by any type of sedation, or even by general anaesthesia. This makes it an especially useful method in evaluating the hearing of small children.

I 2. Basis Principles

I 2.1 Receptor Potentials

Besides the resting potentials (e.g. endolymphatic potential of ± 80 mV re scala vestibuli potential) two stimulus related potentials are present in the cochlea. These will be described in the next paragraphs.

I 2.2 Cochlear Microphonics

The cochlear microphonics (CM), the alternating current cochlear potential, can be recorded from almost anywhere in the cochlea. It is generally accepted, that the CM is produced by the haircells, as is shown in experiments with 'waltzing' guinea pigs which have abnormal haircells due to a congenital defect of the cochlea and the vestibular apparatus. These guinea pigs produce no CM, which indicates that the CM may provide some information about the validity of the haircells (Davis et al., 1934). Von Békésy (1960) found the CM in the normal cochlea proportional to the displacement of the basilar membrane. Dallos et al, (1972 a) showed with experiments in kanamycin-intoxicated guinea pigs that the CM generated by the inner haircells, which appeared at least morphologically normal, is about 30-40 dB less sensitive than the CM generated by outer haircells. This restricts the use of the CM as an indicator for outer haircell validity only.

In promontory recording the CM may give some information about the validity of the haircells in the basal turn, but especially the presence of artefacts which are very difficult to eleminate, makes its clinical usefulness limited.
These artifacts easily arise because the waveform of the CM is similar to that of the stimulus itself. Only a frequency-dependent phase shift occurs. Hence leakages or cross-talk picked up from the electrode or the response recording part of the equipment are not distinguishable from the CM response.

This makes the CM a disturbing feature in clinical electrocochleography. As a routine procedure, the CM is cancelled out from AP en SP recordings by presenting the tonebursts in alternating phase and counterphase, using an electronic commutator.

I 2.3 Summating Potentials

It is generally assumed, that the second stimulus-related potential, the summating potential (SP), is as for the CM, generated near the cuticular lamina of mainly the outer haircells as a result of non-linearities in the movement of the basilar membrane, or non-linearities in the mechano-electrical transduction (Whitfield and Ross, 1965).
The summating potential can easily be studied, since it appears as a DC shift of the baseline which may be either positive or negative with respect to the scala tympani potential (Tasaki etal, 1954). Whenever a summating potential is present, it appears as a part of the waveform (which consists of AP and SP), while the CM is cancelled out by the averaging. Guinea pig experiments with intracochlear electrodes have demonstrated that the SP changes sign between scala media and scala tympani. In promontory recording the sign of the SP is called negative when the DC shift is of the same polarity as the initial part of the compound AP.
For a round window recording in the guinea pig one nearly always records a positive SP when stimulating with high-intensity high-frequency tonebursts. In recording from the round window niche or the promontory in man an SP^- is generally recorded (Eggermont et al., 1974) for all frequencies at high intensity levels. Occasionally an SP^+ is recorded, or a change from negative to positive by increasing the stimulans frequency at constant intensity, but mostly in pathological ears. By further increasing the frequency, the amplitude of the positive summating potential increases simultaneously. It is known from animal experiments, that the sign of the summating potential is not constant along the whole cochlea, but depends on the site of the electrode and the frequency used.
In animal experiments, usually intracochlear electrodes are used, one in the scala vestibuli and one in the scala tympani. A differential (DIF) and overall (AVE) SP can be recorded. The DIF-SP represents the DC shift between scala vestibuli and scala tympani, the AVE-SP the DC shift of the entire cochlea relative to the neck muscle (Dallos et al., 1972b). Only the AVE-SP wil be discussed here, since it closely resembles the summating potential recorded in human promontory electrocochleography, which records the DC shift between the cochlea, or at least its most nearby areas, relative to the earlobe.
Fig 1 shows a scheme of the cochlea with two intracochlear electrodes in the basal turn. Schematically the envelope of the traveling wave is drawn in. The summating potential changes sign at the falling part of the traveling wave (Dallos et al., 1972b). Therefore, these electrodes will record a negative AVE-SP. The lower diagram shows the envelope of the electrical traveling wave from a stimulus with a higher frequency. The falling part of the envelope is now located more near the basis of the cochlea, and a positive AVE-SP will be recorded. Since in electrocochleography in man the electrode is placed on the promotory near the basis of the cochlea, the recorded SP is nearly always negative, because the electrode is very near the rising part of the traveling wave envelope.

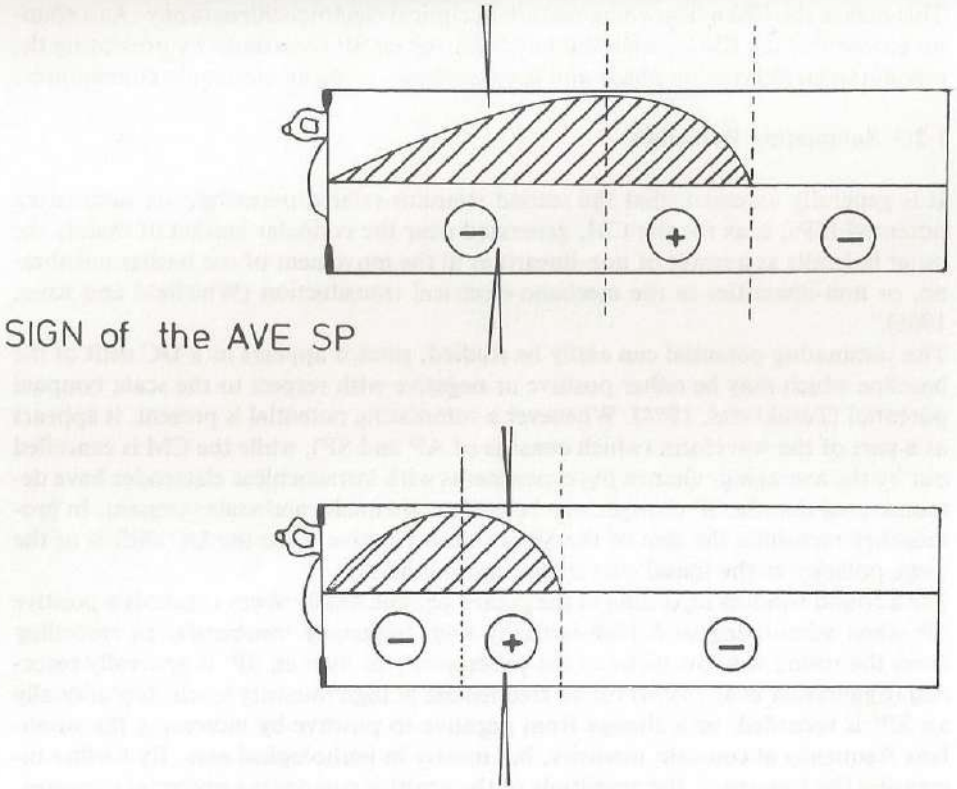


Fig. 1 The cochlea and the envelope of the electrical travelling wave with two intracochlear electrodes are schematically represented.
In the upper diagram a negative SP will be recorded. In the lower a positive SP.
This is the result of different electrode locations and a different stimulus-frequency.

I 2.4 The Neural Response: the compound action potential

In the cat, 90-95% of the afferent fibres of the auditory nerve are radial fibres innervating the inner haircells, leaving only 5-10% of spiral nerve fibres innervating the outer haircells (Spoendlin, 1970). In guinea pigs almost the same distribution exists (Morrison et al, 1975); therefore it is assumed that the percentages will be the same in man. Consequently, the majority of recordings from single nerve fibres are from radial nerve fibres. Since the compound action potential (AP) is an extracellularly representation of the activity of the bundle of individual neurons (the nerve), the contributions of single nerve fibres to the compound AP will be primarily from fibres innervating the inner haircells.
In promontory recording with the reference electrode at the earlobe, a diphasic AP is usually obtained. The amount of synchronization of the individual nerve fibres is

very important for the shape of the compound AP and depends on the stimulus frequency. For low frequencies (< 2000 Hz) a phase lock appears and an AP is recorded on each cycle of the sinewave. Due to adaptation and refractoriness, the AP amplitude diminishes with the successive number of the period. Above 2000 Hz, the phase lock disappears for the compound AP and only the response to the start of the stimulus is recorded (fig. 2). This is called the 'on-effect'.

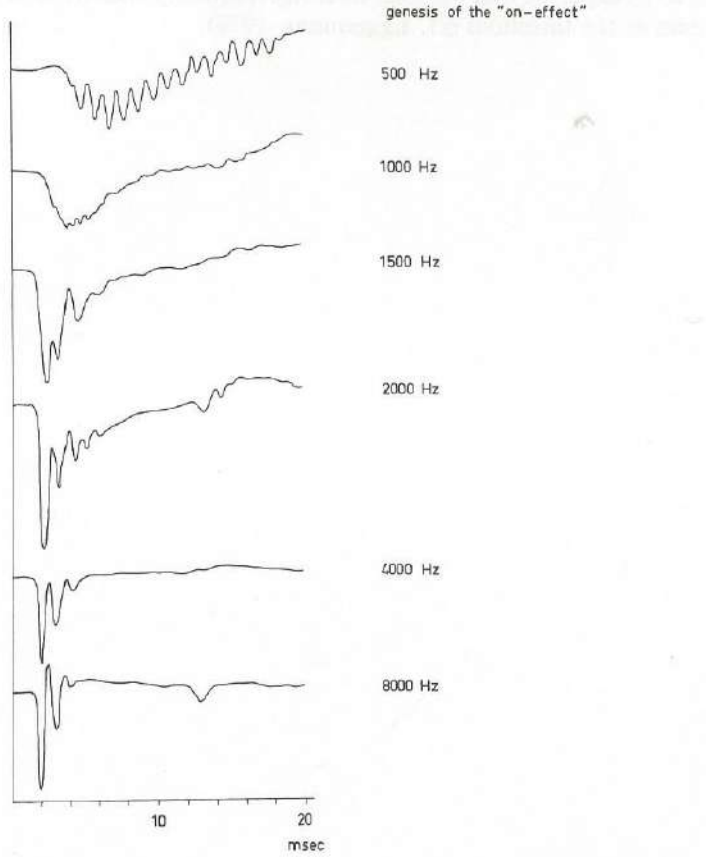


Fig. 2 Genesis of the 'on' effect. (guinea pig)
In response to tone-bursts having two periods of the sinewave during the rise- and fall time and six periods during the plateau, the shape of the compound AP depends on the stimulus frequency. For low frequencies (< 2000 Hz) a phase lock appears and an AP is recorded on each cycle of the sine-wave.
Above 2000 Hz the phase lock disappears for the compound AP and only the response to the start of the stimulus is recorded. All the recordings were made at intensities of 60 dB SPL. (from Eggermont et al., 1974).

Eggermont et al. (1974) demonstrated the possibility of frequency selective threshold determination with the use of frequency specific tonebursts. These tonebursts have a constant number of cycles, but different rise and fall times and plateau duration. This will not be discussed in detail. With increasing stimulus intensity the AP latency decreases, the latency shift being largest for lower frequencies. At high stimulus intensities the AP latencies are nearly the same for tone-bursts of frequencies from 2 kHz to 12 kHz, but at threshold values the latency depends on the tone frequency. This increase in latency for the whole nerve response is mainly due tot an apical shift of the excitation along the cochlear partition. Because the synaptic delay is not frequency dependent, one can say that at threshold values the excitation areas are clearly separated, but at higher intensities tend to overlap basally. Therefore, it is to be expected that the high-intensity frequency specificity of AP recordings is less than at the threshold (cf. Eggermont, 1976).

II Methods

II 1.1 Electrodes

The transtympanic electrode technique is commonly used nowadays. A 01 mm diameter electrode is placed on the promontory through the pierced eardum (fig. 3). Different placements of the electrode (e.g. ear lobe, ear canal) result in much smaller amplitudes of the recorded potentials. (Eggermont et al, 1974).
All electrocochleographic studies in Leyden are performed by using this transtympanic electrode as the active electrode. The reference electrode is placed at the ipsilateral mastoid or ear lobe, the ground electrode at the forehead.

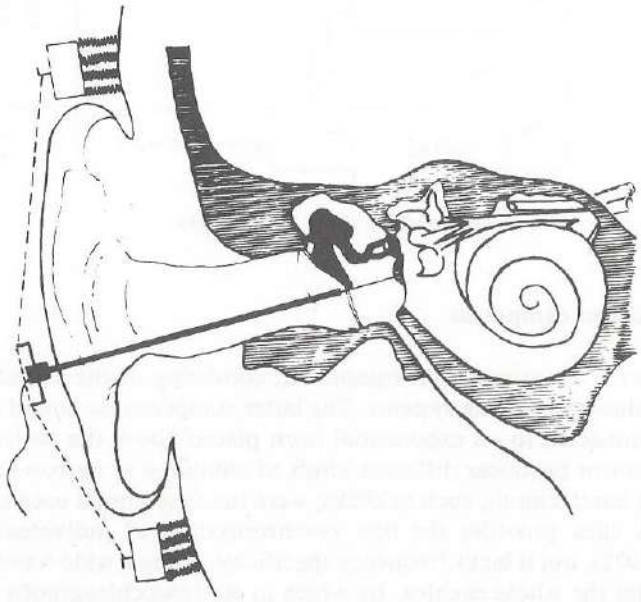


Fig. 3 Transtympanic recording (from Eggermont et al., 1974).

II 1.2 Recording system

The apparatus used in electrocochleography in Leyden (Spoor, 1974) consists of two parts, one being the recording system consisting of the preamplifier and the main

amplifier (to magnify the weak biological signals to adequate magnitude) plus an oscilloscope to observe, and an instrumentation tape recorder to record the amplified signal. This signal is also fed to an electronic averager plus X-Y recorder to enhance the relevant signal with respect to the back-ground and to store it on paper (Fig. 4).

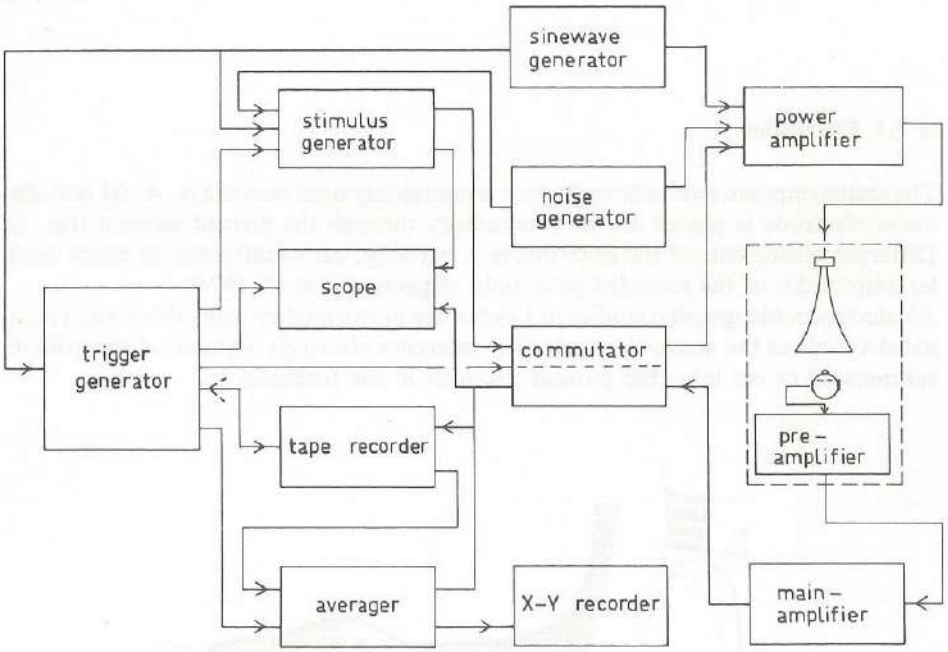


Fig. 4 Block-diagram of the recording system (from Spoor, 1974).

II 1.3 Stimulation equipment

The other part is the stimulation equipment, consisting of the stimulus-generating and the stimulus-transducing systems. The latter comprises the sound amplifier plus driver unit connected to an exponential horn placed above the patients head. The stimulus generator produces different kinds of stimuli, e.g. narrow- and wideband stimuli. Wide band stimuli, such as clicks, were the first stimuli used in electrocochleography. A click provides the best synchronization of individual nerve fibers (Aran et al, 1972), but it lacks frequency specificity. Being a wide-band stimulus, the click stimulates the whole cochlea, by which in electrocochleography a general impression of cochlear function can be obtained. By filtering the clicks using a band-pass filter, a frequency specific character can be given to the click, especially by using 1/3 octave filtered clicks. This improves frequency selectivity in threshold determination (Aran et al, 1971). Another frequency specific sound stimulus was first used by Yoshie (1971), and later by Eggermont et al. (1974). These trapezoïdalshaped tonebursts have subsequently been used for all the electrocochleograms made in Leyden.

The pure-tone bursts have two periods of the sinewave during the rise-time and the fall-time and at least six periods during the plateau. This makes the duration of the toneburst dependent of the stimulus frequency. The shortest rise time, however, is 0.33 msec and the plateau duration is never taken less than 4 msec. to assure reliable SP measurements.
Using tone-bursts, it is important to cancel the CM. To archieve this, the tonebursts are presented alternately in phase and counterphase by using an electronic commutator.

II 1.4 Duration, anaesthesia, premedication

The whole electrocochleographic procedure studying one ear for 500, 1000, 2000, 4000 and 8000 Hz takes one to one and a half our. Unless it concerns a small child, the examination will be carried out in local anaeasthesia and nowadays even without local anaesthesia. The local anaesthesia is given by infiltrating the ear canal with 1cc xylocaine 2% combined with adrenalin 1:80.000.
Since it is necessary for accurate recordings that the patient lies motionless, a sedation is given. For an adult patient this premedication consists of 150 mg Nembutal® orally one and a half hour before ECoG and 50 mg pethidine and 25 mg Phenergan® intramuscularly a half hour before ECoG. Usually the patients sleep during the whole procedure.
If the indication to perform electrocochleography is strong, while it is not possible to use a local anaesthetic (e.g. multihandicapped children or children with autistic behaviour) the procedure can be performed under general anaesthesia.

II 2 Practical diagnostic procedure

II 2.1 Introduction

When the cochlea is stimulated with tone-bursts presented alternately in phase, it is important to realize that phaselock between the sinewave and the single nerve AP's occurs for frequencies below 1500 Hz. For 500 and 1000 Hz alternation of the stimulus-phase in order to abolish the CM, leads to a half period shift of the preferred phase. This in turn causes a doubling of the number of AP's with respect to the sine period. These closely spaced AP's overlap partially, thus forming a broad average response in which only the AP-tips indicate the phaselock (Fig. 2).

II 2.2 Threshold determination

Thresholds are routinely determined for frequencies from 500 to 8000 Hz. Hearing threshold is fixed at an AP amplitude of 0.1 μV or less (fig. A.1.1 lower right, page 28). Spoor and Eggermont (1976) studied approximately 100 patients from which a reliable pure tone audiogram was available, and 60 young children from which it was possible to get a fair impression of their hearing abilities with observation or free field audiometry. Their observations justify the conclusion that threshold determination with toneburst-ECoG is just as reliable as with pure-tone audiometry.

II 2.3 Amplitude-intensity relationships

AP amplitude values are calculated from the baseline to the first peak (N_1) of the compound AP. When AP amplitudes are plotted on a logarithmic scale, and the corresponding intensity values on an linear scale, an amplitude-intensity curve can be drawn. These relationships are of major importance in studying supraliminal hearing characteristics.

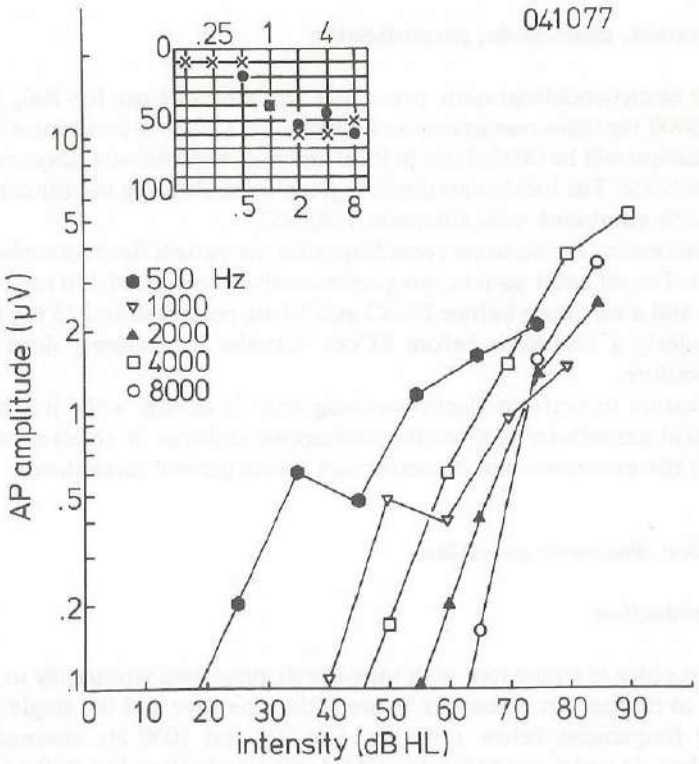


Fig. 5 Amplitude-intensity curve. The steepness of the curves increases with increasing frequency. At intensities of 50-60 dB a plateau in the curves is seen.

An example of such a set of curves is shown in Fig. 5. In general, the steepness of the curves increases with increasing frequency. Especially at 50-60 dB intensity levels, a plateau in the curve is often seen, the origin of which is still subject to discussion (Davis, 1976). This plateau divides the curve in an low (L) and high (H) part (Yoshie, 1968). This phenomenon is more clear at lower frequencies.
The output of normal ears differs largely and therefore cannot be used as a parameter. In a group of 20 normal ears Eggermont (1976) found a 10-fold amplitude range (2-20 μV) at 85 dB in case of 2000 Hz stimulation.
Comparison of atlas A.1.1 (page 28) and B.2.12 (page 64) results in a factor 2-3 difference at high-output levels for two normal hearing ears, while for example the

maximum output in a pathological ear with 40 dB perceptive loss (atlas B.1.11, page 60) may vary between 10 en 20 microvolts.

II 2.4 Latency-intensity relationships

From the AP responses shown in Fig. A.1.2 right below (page 30), it can be observed that the AP latency, calculated from CM onset to the largest AP peak, increases with decreasing stimulus intensity. If the logarithm of latency is plotted as a function of the stimulus intensity, one usually obtains a nearly linear relationship in case of toneburst stimulation. The curves for different frequencies are nearly parallel. Sometimes, however, the AP shows a double peaked waveform at intensities of approximately 50 dB, which is related to the L and H part of the amplitude-intensity-curve. Since, by convention, the largest peak is chosen for latency calculations, this may cause a relatively large shift in the latency-intensity curve.

II 2.5 Amplitude-latency relationships

When the AP amplitude is plotted versus AP latency on a double logarithmic scale, a mostly linear relationship results. By extrapolation to the X-axis, the latency at threshold can be calculated for different frequencies. The normal hearing characteristics of the amplitude-latency curve have been described (Eggermont et al, 1974). It appears that for conductive hearing loss the data are within the normal range, but for the majority of the cochlear hearing losses the lower amplitude values are outside the normal range.

II 3 Scope of the investigation

1. A study of the clinical use of the summating potential in electrocochleography.
2. Demonstration of 20 selected cases to illustrate and evaluate the various outcomes of the test.
3. Investigation into the actual influence of electrocochleography on the management of the hearing disorders in about 500 cases.

III 1. SP-Polarity

For high frequency, high intensity stimulation in man, generally a negative summating potential is recorded. In some pathological cochleas, however, a positive summating potential (SP^+) is seen (Aran et al., 1971a; Eggermont et al., 1974; Eggermont, 1976; Nishida et al., 1976).
The relationship of the SP polarity to the shape of the audiogram will be investigated and a model explaining the observation will be proposed.
In about 25 human ears an SP^+ has been observed at one or more stimulus frequencies. For a fixed stimulus intensity the SP^- change to SP^+ was always observed for increasing frequency. In the introduction (I.2.3.) it has been argued that a change in SP polarity can be observed in normal guinea pig cochleas using the differential electrode technique by a sufficient change of stimulus frequency. In the promontory recording the SP will always be negative for a normal functioning cochlea (Fig. 6, upper trace).
In case the most basal past of the cochlea is devoid of hair cells (e.g. as a result of ototoxic damage), part of the activity pattern which normally produces an SP^- is missing. The result will be a reduced SP^- amplitude (Fig. 6, second trace). If under these conditions a higher stimulus frequency is used, resulting in a basal shift of the excitation area, the SP^- producing part, which is normally dominating, may be compensated by the SP^+ producing part. The result could be a very small or absence of SP (third trace).
A further increase in stimulus frequency may cause only activation of the SP^+ producing part of the excitation pattern, resulting in an SP^+ recording at the promontory.
According to this, it can be stated that a positive SP will be recorded for high frequencies only, when hair cell damage is located near the basis of the cochlea. If hair cell damage also spreads to more apical parts of the cochlea, positive summating potentials will be recorded for the lower frequencies too. (Rietema and Eggermont, 1976).
In Fig. 7 auditory thresholds and mean audiograms are shown for those ears in which an SP^+ at one or more frequencies was recorded. The ears are grouped according to an SP^+ found only at 8 kHz, at 4 and 8 kHz, and at 2,4 and 8 kHz. From the survey of these data the conclusion can be drawn that the occurrance of SP^+ at lower frequencies generally requires an increase in the amount of hearing loss which is especially obvious for the third group. The notion that recording of SP^+ in patients suffering from sudden deafness was pointing to a bad prognosis (as to its recovery, Nishida et al., 1976) seems to fit with the above mentioned model.

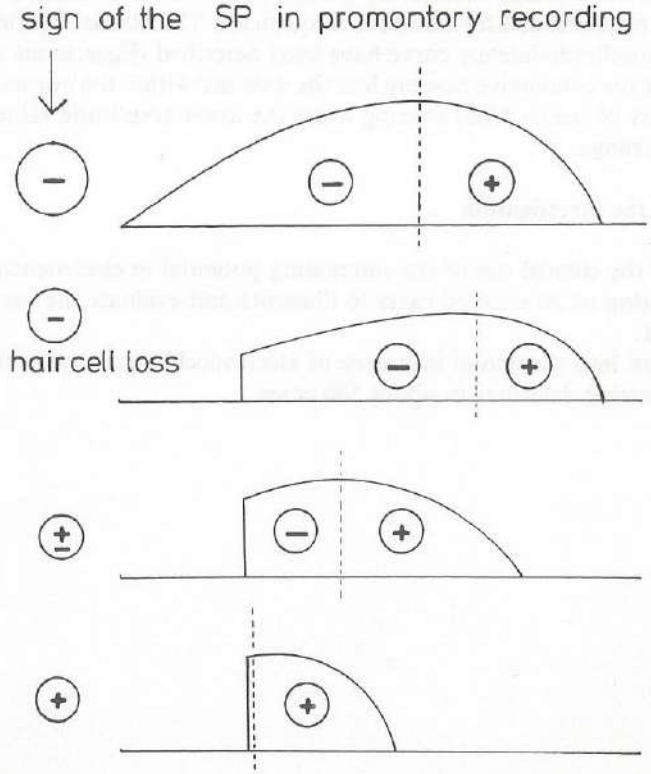


Fig. 6 Schematic representation of the envelope of the electrical travelling wave in a normal cochlea (upper tracing) and in a pathological cochlea for different frequencies (lower tracings). Frequency increases from right to left. (Note that the amount of hair-cell loss in the lower three tracings is not changed)

III 2 SP amplitude

The promontory recorded average summating potential amplitudes at 85 dB are schematically drawn in in fig. 8.
Also one standarddeviation upwards is indicated for 25 normal ears (open circles), 25 Menière ears (squares) 16 hair cell loss ears (triangles) and for a patient with a sensorineural loss due to secondary syphilis (see also case nr B. 3.15, page 75).
The hair cell loss ears are those with a history of noise trauma or the use of ototoxis drugs.
In general, there is not much difference between the SP amplitudes of Menière ears and normal ears for 2, 4 and 8 kHz at this stimulus level. However, the hair cell loss ears produce a smaller SP or even no SP, which is understandable since the SP is produced by normal functioning hair cells. On the other hand, it can be concluded that there is no indication for hair cell loss in the Menière group.
It is also noted that the SP amplitude increases for higher stimulus frequencies, which is also found is hair cell loss ears. However, in Menière ears the larger SP^- va-

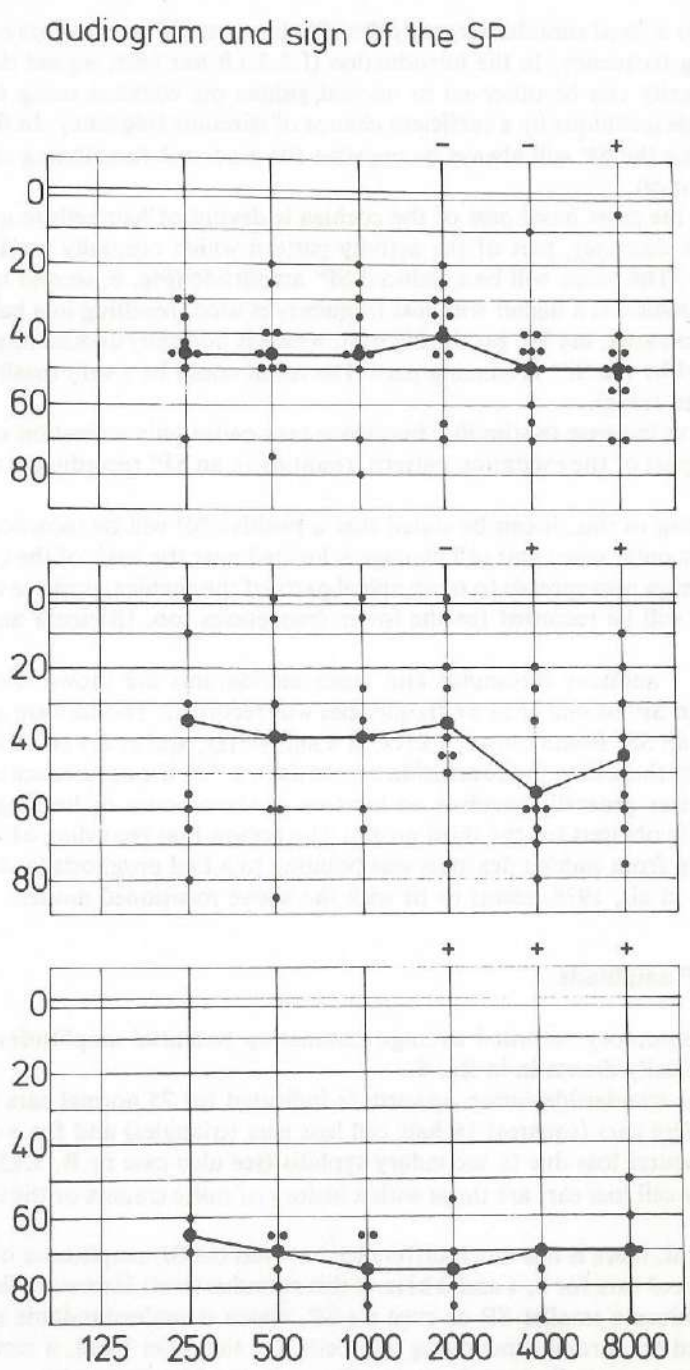


Fig. 7 Three groups of audiograms for which a positive SP is recorded for one or more frequencies.

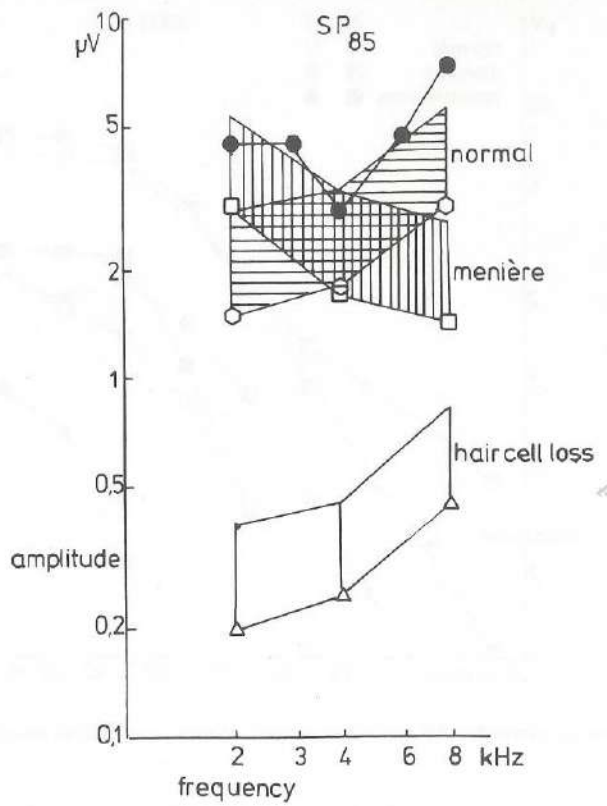


Fig. 8 Average Summating potentials in man.
open circles : normal cochleas
triangles : haircell loss
rectangles : Menière's disease
closed circle : Secondary syphilis (case B.3.15)
The areas above the curves represent one standard deviation.

lues are found for the lower frequency.
The input-output properties of the SP as well as the AP are plotted in fig. 9. The results are taken from the same group of normal, Menière and hair cell loss ears, thusfar described. For the hair cell loss ears the average AP threshold is 55 dB HL and the average SP detection threshold is shifted by about 20 dB with respect to those of normal ears. For the Menière ears the mean AP input-outputcurve closely follows that for normal ears, but for intensity values below 60 dD HL falls progressively below the normal curve. The average thresholdshift for the Menière ears is about 25 dB for 2000 Hz, but the detection threshold for the SP in these Menière ears is 15 dB lower than for normal ears.
The SP is supposedly produced as a result of an asymmetry in the movement of the basilar membrane. If, due to endolymphatic hydrops, which is assumed to be pre-

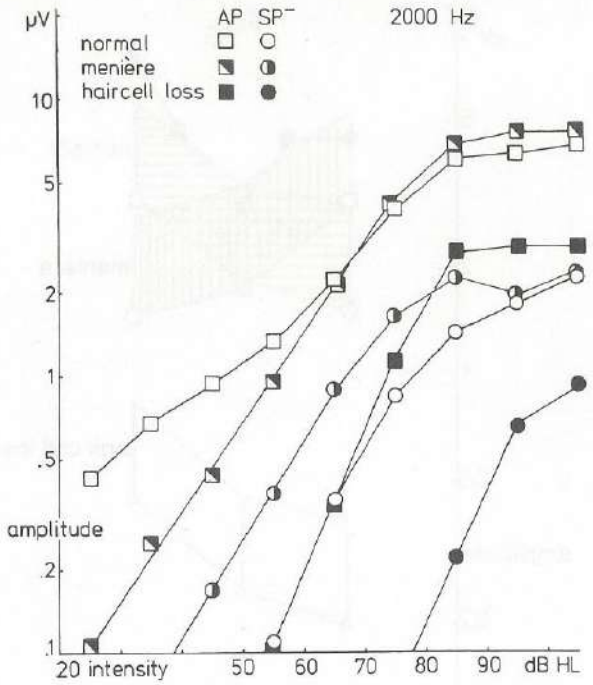


Fig. 9 Input-output curves for AP and SP in normal, Menière and haircell loss ears (from Eggermont, 1976).

sent in Menière ears, a static displacement of the basilar membrane is already present, the asymmetry of the movement of the basilar membrane will increase. This might explain the smaller detection threshold for the SP in Menière ears and an enlarged amplitude of the SP (Johnstone, 1975). This implies that the SP tentatively might be considered as the detector of endolymphatic hydrops (Eggermont, 1976). An electrocochleographic study of the effects of glycerol administration in 13 Menière patients supports this theory, the intake of glycerol resulted in nine patients in a decrease of SP amplitude (Moffat et al., 1978).

Summary

Because the SP is less likely to be produced by artificial signals than the CM, the use of the SP for diagnostic purposes is considered more valid. The presence of a positive SP or a small or absent negative SP for various frequencies points to hair cell loss, in Menière ears the negative SP found for 2000 Hz appears to be normal. The input-output properties of the SP^- are important in differentiating early stages of Menière's disease from hearing loss due to other cochlear lesions.

IV Electrocochleographic characteristics of several types of hearing loss

Different types of hearing loss show different electrocochleographic features above threshold.
This section, in which the general characteristics of several types of hearing loss are described, may be regarded as an introduction to section V. Section V comprises an electrocochleographic study of 20 selected cases out of about 500 ECoG's performed in Leyden.

IV 1 Conductive hearing loss

As may be expected from a situation with a normal cochlea but impaired middle-ear transmission, the stimulus used will be less effective than in normal ears. Therefore the amplitude-intensity curve is shifted to the right as compared with normal ears (A.1.2. upper right, page 30). This is caused by the thresholdshift only, and the shape of the amplitude-intensity curve is the same as for normal ears. The latency-intensity curves are in general not much different from those found in normal ears; however, a tendency to longer latencies seems to exist (A.1.2. lower left, page 30).
Summating potential amplitudes in conductive hearing loss tend to be smaller than in normal ears, but in most cases the large standard deviation for SP amplitudes does not justify a conclusion.
Finally, the AP waveform cannot be distinguished from those found in normal ears either, so that AP threshold elevation remains the most important characteristic feature of a conductive hearing loss.

IV 2 Sensorineural hearing loss

In case of sensorineural hearing loss with recruitment, the relationships for the AP amplitudes and AP latencies differ fundamentally from those of the normal cochlea.
An example of amplitude-intensity curves in a recruiting ear is given in fig. B.1.9 upper right (page 52).
Although threshold values are elevated, amplitudes reach high-intensity values which are the same as in normal ears. Just above threshold a rapid increase of amplitudes is observed. The slope of the curve in recruiting ears is much steeper than in normal ears. The latency-intensity relations at high intensity values are comparable to those for normal ears; in cases of high-frequency sensorineural hearing loss the latency near threshold tends to be shorter than normal (atlas B.1.10 lower right, page 56).

A distinction between conductive and sensorineural hearing loss is made on basis of a) threshold values, b) the slope of the amplitude-intensity curve, c) the slope of the amplitude-latency curve and d) latency values near threshold, (Hermans et al., 1975).
Distinction between Menière's disease and other types of sensorineural hearing loss is made on basis of the characteristic AP waveform in Menière's disease, which is caused by a relatively large SP^- part of the compound AP (Schmidt et al., 1974). Small SP amplitudes and/or positive polarity are indicative for outer-haircell loss (cf. section III). When a mixed hearing loss is present (A.2.6. upper left, page 41) interpretation of ECoG results becomes more complicated and only tentative results can be expected.

IV 3 Hearing loss due to acoustic neuromas and central lesions

A distorted and relatively broad waveform is the most characteristic feature in acoustic neuromas. This is reported by many authors (e.g. Eggermont, 1976). An example of such a typical waveform is given in B.2.13 lower right (page 67).
At intensities of about 60 dB the N_1 disappears in favor of an N_2 which becomes more prominent. This phenomenon is reflected in the amplitude-latency function by a rather abrupt latency shift (B.2.13 lower left, page 67), which is never seen in normal ears or in conductive hearing loss. It must be noted (as in B.2.13, page 66) that in pontine angle neuromas subjective hearing may be absent, while normal thresholds and amplitude intensity curves may be found in electrocochleography.
This difference (in variable degrees) between subjective hearing thresholds and ECoG thresholds is also present in other types of central hearing loss (located central to the pia-mater) (B.2.14, page 70). In B.3.16, page 78, a partially cochlear, partially central hearing loss is seen.
Often, a rather flat amplitude-intensity curve is seen in these cases, but apart from the threshold difference no specific electrocochleographic pattern is found. Brainstem audiometry probably may provide more information in these cases (Selters and Brackmann, 1976).

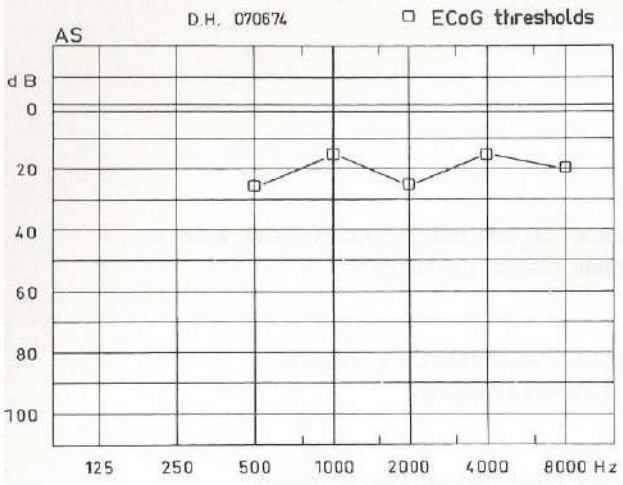
V Clinical results II: Survey of selected ECoG cases

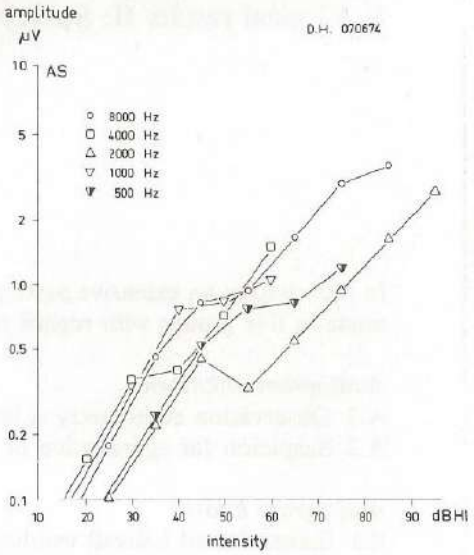
In this chapter an extensive description of 20 selected patients is given; a division is made in five groups with regard to their clinical problems:

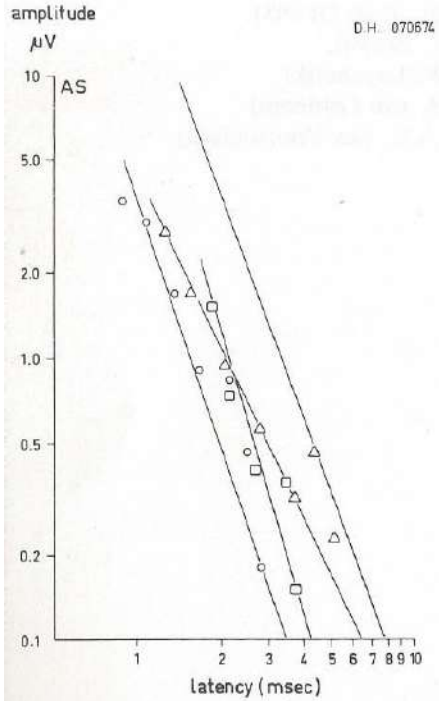
Audiogram unknown
A.1 Observation audiometry is impossible or unreliable (5 cases).
A.2 Suspicion for aggravation or simulation (3 cases).

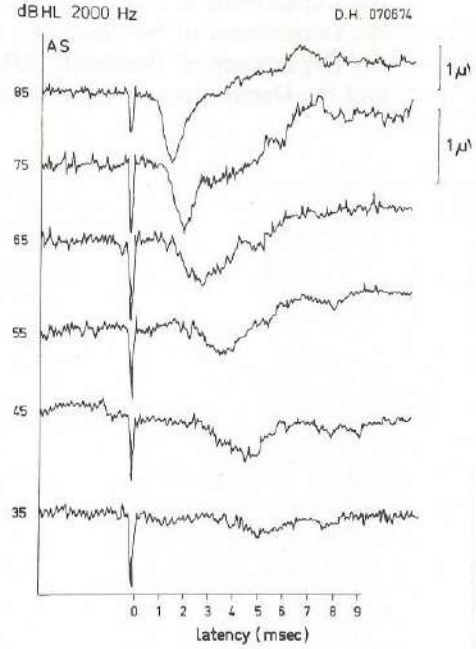
Audiogram known
B.1 Estimation of haircell validity in cochlear hearing loss (3 cases).
B.2 Diagnosing retrocochlear or central types of hearing loss (3 cases).
B.3 Predicting the prognosis in sudden deafness (5 cases).

Anaesthesiological, internal, neurogical, neurosurgical, pediatric and radiological consultations were carried out by
the Department of Anaesthesiology (Head: Prof. dr. Joh. Spierdijk),
the Department of Internal Medicine (Head: Prof. dr. J. de Graeff),
the Department of Neurology (Head: Prof. dr. G.W. Bruyn),
the Department of Neurosurgery (Head: Prof. dr. W. Luyendijk),
the Department of Paediatrics (Head: Prof. dr. H.H. van Gelderen)
and the Department of Radiology (Head: Prof. dr. A.E. van Voorthuisen).









A 1.1 Autistic child with normal hearing

Case nr. 070674

History

A fourteen months old girl was hospitalized in our clinic for hearing evaluation.
When the child was nine months old the mother became convinced that she was hard of hearing.
The girl was born after a pregnancy of 38 weeks, which had been complicated by a pyelonefritis of the mother for which she was treated with ampicillin. Alupent® intravenously was given during two weeks to prevent immature delivery.

Examinations

Normal eardrums and normal tubal functioning were found. A pediatrician was consulted, who found motoric retardation and suspected a mental retardation too.

Audiometry

Observation audiometry was performed twice. Except for the sound of an autoclaxon (approx. 100 dB SPL) no reaction on standard sounds was observed.
It was noticed that the girl was heavily involved with parts of her own body: fingers, lips, teeth etc. It was not possible to get eye to eye communication with the child.

Indications for electrocochleography

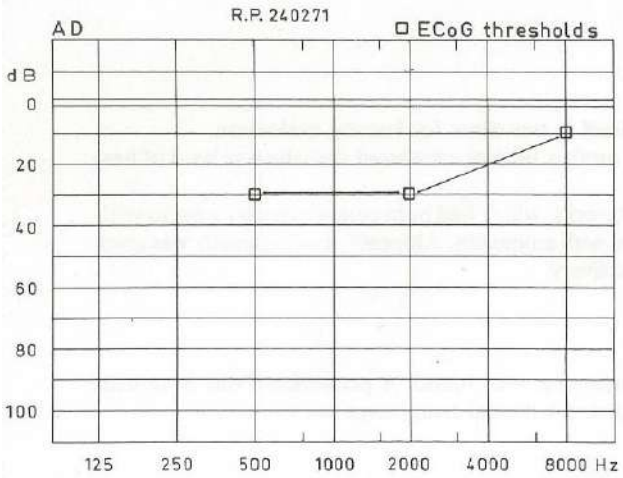
It was clear that the child was motorically retarded.
With observation audiometry it was not possible to get useful results with respect to hearing ability. Therefore ECoG was indicated to differentiate between mental retardation of unknown origin, and a retardation due to a congenital hearing loss.

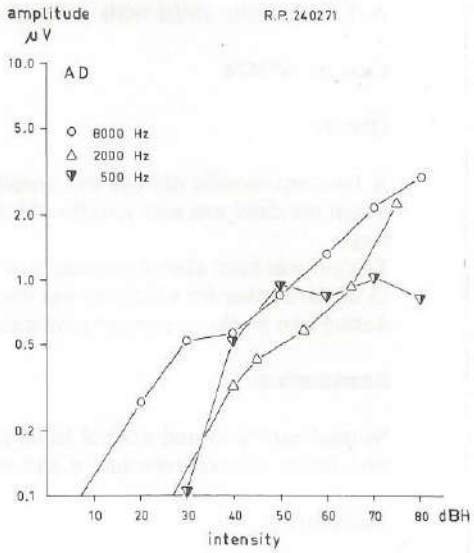
Results of electrocochleography

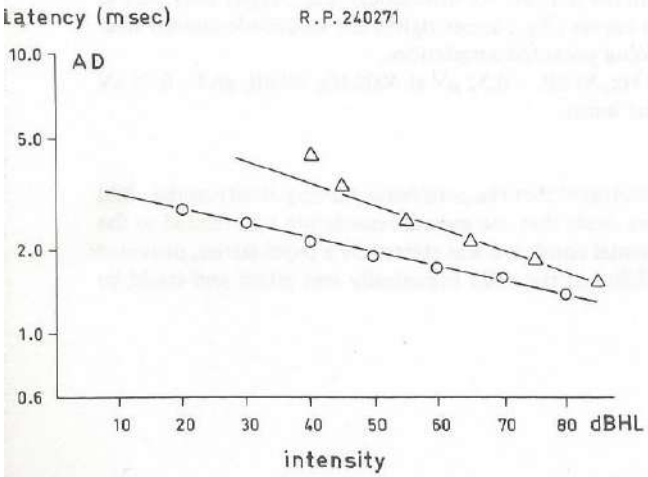
Electrocochleography was performed on the left ear. AP-thresholds (Fig., upper left) were in the normal range, as were input-output curves (Fig., upper right), the amplitude-latency relationships (Fig., lower left) and summating potential amplitudes.
SP amplitudes were: $-0{,}56\,\mu V$, at 2000 Hz, 85 dB; $-0{,}52\,\mu V$ at 4000 Hz, 80 dB; and $-0{,}75\,\mu V$ at 8000 Hz, 85 dB, being within normal limits.

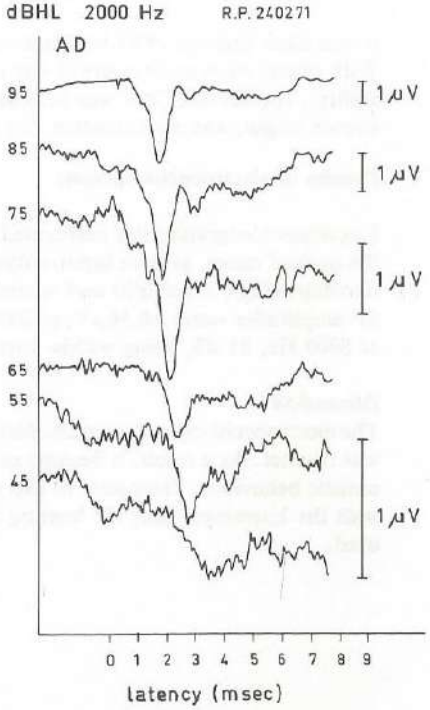
Discussion

The electrocochleographic results demonstrated that the peripheral hearing ability of the child was normal. As a result, it became more likely that the mental retardation was related to the autistic behaviour. Treatment of this mental condition was started by a psychiatrist, provided with the knowlegde that the hearing ability of the child organically was intact and could be used.









A 1.2 Conductive loss due to glue in the middle ear

Case nr. 241271

History

A two months old boy was referred to our clinic by his family doctor. The boy's mother questioned his ability to hear, which was understandable because two other children in this family suffered from progressive hearing loss, after having been hearing normal at first. They both became hard of hearing at the age of 4 and 7 months respectively.

Examinations

Normal eardrums were found.

Audiometry

Observation audiometry showed clear reactions on tones for the whole frequency range. This audiometry was repeated every two months. At the child's age of six months his parents believed that hearing had become worse. This time observation audiometry showed a moderate to severe hearing loss. Free field slow-vertex response audiometry showed a 50 dB threshold for 1000 and 4000 Hz.

Indications for electrocochleography

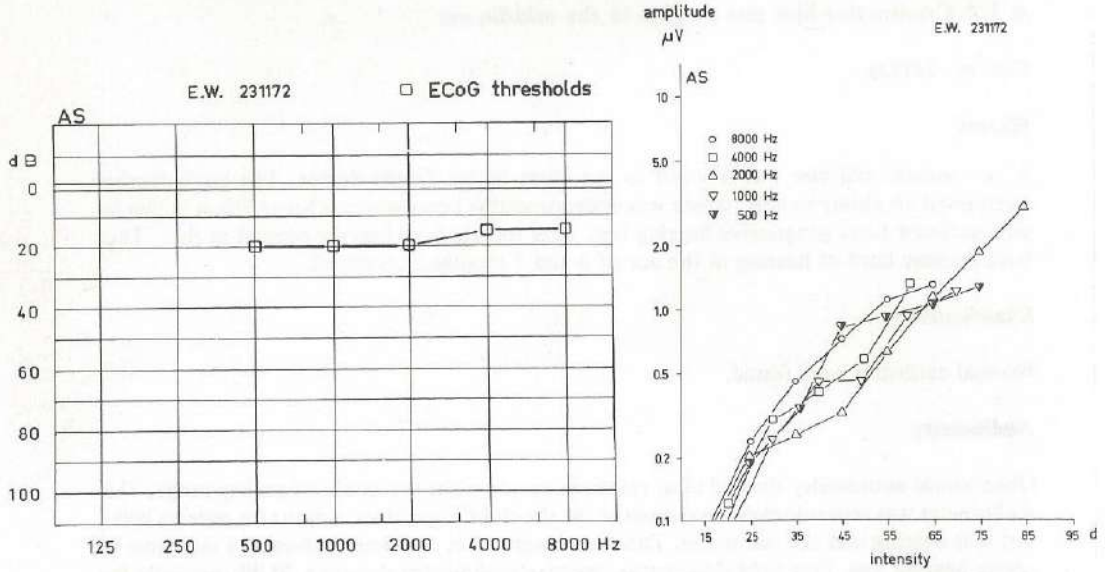
Since a considerable hearing loss was suspected, ECoG was indicated to determine hearing thresholds, and in case of a hearing loss, to discriminate between a conductive and a perceptive type.

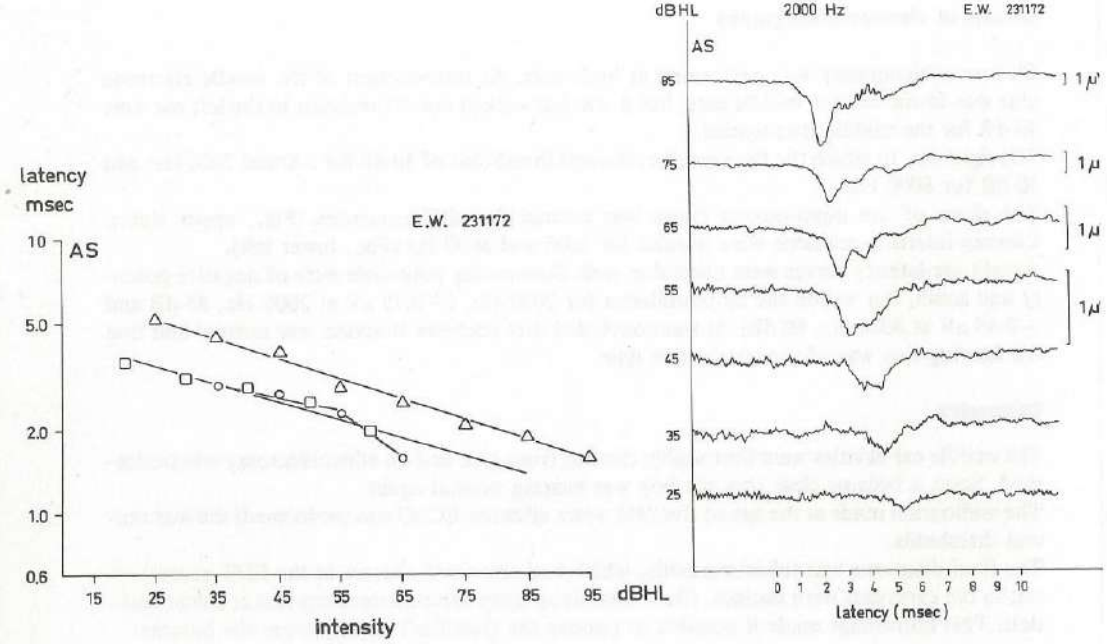
Results of electrocochleography

Electrocochleography was performed at both ears. At introduction of the needle electrode glue was found in both middle ears, but it was not sucked out. Thresholds in the left ear were 40 dB for the middle frequencies.
The right ear, to which the figures refer, showed thresholds of 30 dB for 500 and 2000 Hz, and 10 dB for 8000 Hz.
The slope of the input-output curve was normal for all frequencies (Fig., upper right). Latency-intensity relations were normal for 2000 and 8000 Hz (Fig., lower left).
Amplitude-latency curves were normal as well. Summating potentials were of negative polarity and small, but within the 2σ boundaries for 2000 Hz. ($-0.75\ \mu V$ at 2000 Hz, 85 dB and $-0.65\ \mu V$ at 8000 Hz, 80 dB). It was concluded that cochlear function was normal and that the hearing loss was of the conductive type.

Discussion

The middle ear cavities were thoroughly cleaned from glue and an adenoidectomy was performed. Soon it became clear that the boy was hearing normal again.
The audiogram made at the age of five (4½ years after the ECoG was performed) showed normal thresholds.
The final diagnosis was tubotympanitis, which had remained obscure at the ENT examination, as the eardrums were normal. Electrocochleography demonstrated normal cochlear function. This knowledge made it possible to remove the (justified) anxiety from the parents.





A 1.3 Evaluation of a baby's hearing before gentamycin application

Case nr. 231172

History

A two and a half month old boy had been hospitalized in the pediatrics department since birth. Directly after birth be had been blue and very weak. He had been nursed in an incubator and had not reacted to stimuli during his first weeks. He had developed chronic infection of the urinary system which had been treated unsuccesfully with several antibiotics. The pediatrician considered starting treatment with gentamycin.
Before starting therapy with an ototoxic antibiotic in our hospital it is routine to make an audiogram, which is frequently repeated during therapy. For this reason the ENT department was consulted.

Examinations

Routine ENT examination demonstrated normal eardrums.

Audiometry

An effort was made to perform observation audiometry at this very young age, but the only result was evidence of some hearing.

Indications for electrocochleography

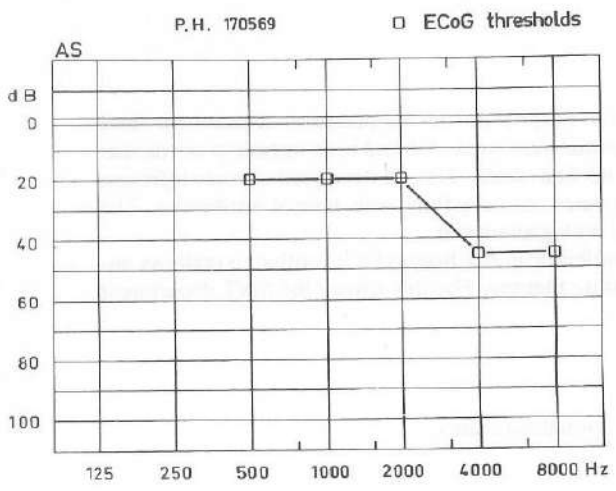
Since it was important to know the exact cochlear function before starting gentamycin therapy, electrocochleography was indicated because no impression of hearing thresholds could be gained by observation audiometry.

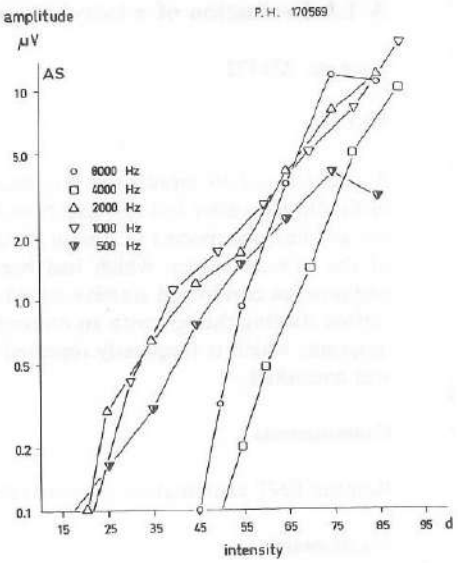
Results of electrocochleography

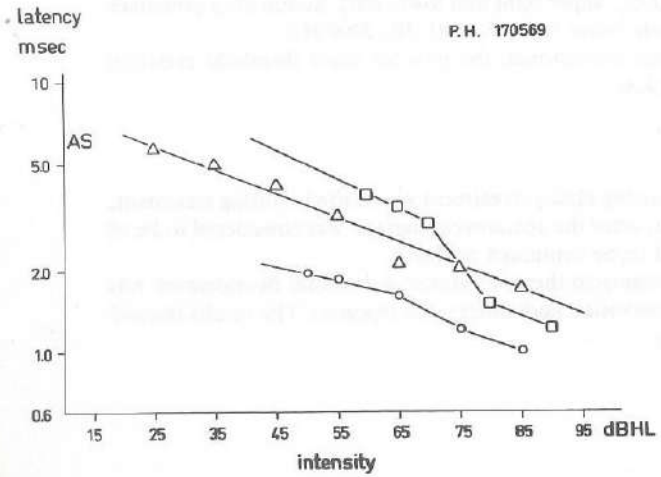
Electrocochleography was performed on the right ear, which showed a normal middle ear. Thresholds were 20 dB for the speech frequencies (Fig., upper left). Input-output curves and latency-intensity curves were normal (Fig., upper right and lower left). Summating potentials were of negative polarity, the amplitude being 1.75 μV at 85 dB, 2000 Hz.
It was concluded that cochlear function was normal; the possible slight threshold evelation might be due to a conductive hearing loss.

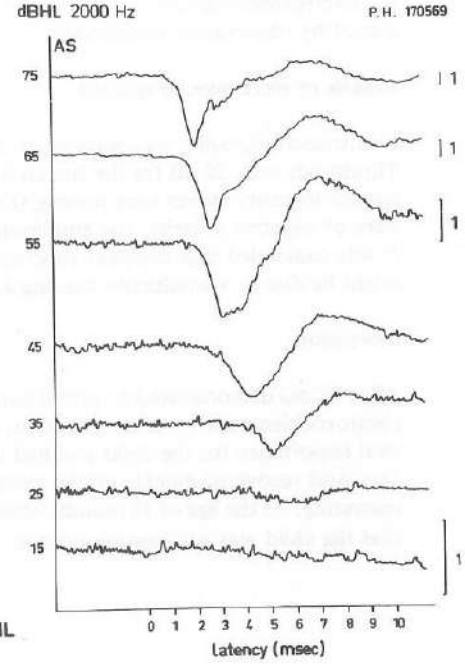
Discussion

After ECoG demonstrated a normal hearing ability, treatment was started. During treatment, electrocochleography was not repeated, since the gentamycin therapy was considered to be of vital importance for the child and had to be continued anyhow.
The child recovered quickly on the gentamycin therapy. Motor and mental development was reassuring. At the age of 10 months observation audiometry was repeated. The results showed that the child was still hearing normal.









A 1.4 A child with Down's syndrome with a mixed hearing loss

Case nr. 170569

History

A 4 year old boy with Down's syndrome was sent to our clinic by the audiologist of a school for deaf children. The child was examined at that school, because it occurred to his present teachers of a school for handicapped children that he often did not react to sounds. At the center for deaf children observation audiometry was performed several times, indicating a conductive loss of 50-60dB, although this could not be stated with certainty, due to his mental retardation. The audiologist of the school requested electrocochleography, in order to obtain more reliable information of the nature of the hearing loss.

Examinations

Strongly retracted eardrums, due to tubotympanitis, were found. Tonsillectomy and adenoidectomy had been performed in the past. The child did not speak. A general physical and neurological examination showed serious psychomotoric retardation. The tendon-reflexes were very low.

Audiometry

Observation audiometry showed good reactions on loud standard sounds at 2 meters distance (about 100 dB HL), for frequencies up to 2000 Hz. It seemed that reactions were worse for sounds above 2000 Hz.
Impedance audiometry was not possible, because the boy was not coöperative.

Indications for electrocochleography

Observation audiometry indicated a hearing loss for both ears,but as mentioned above it was not possible to distinguish between a conductive, perceptive or a mixed loss. The aspect of the eardrums, however, suggested at least a conductive component. For the final decision of placement in either an institute for psychomotoric retarded children or in an institute for multihandicapped children where an eventually existing hearing loss could also be given attention, it was important to know the exact hearing thresholds and the cause of the hearing loss.

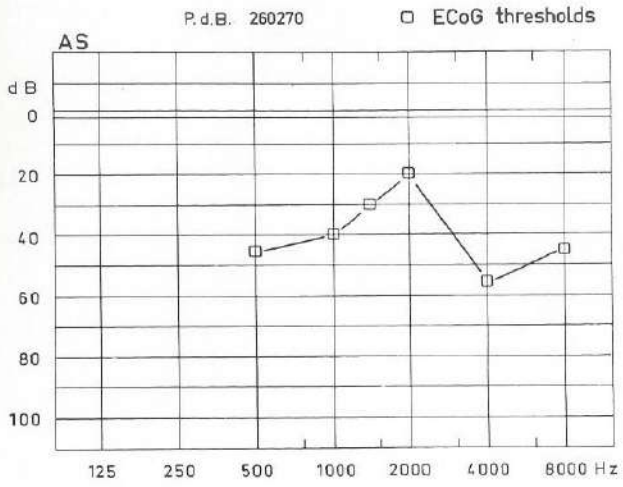
Results of electrocochleography

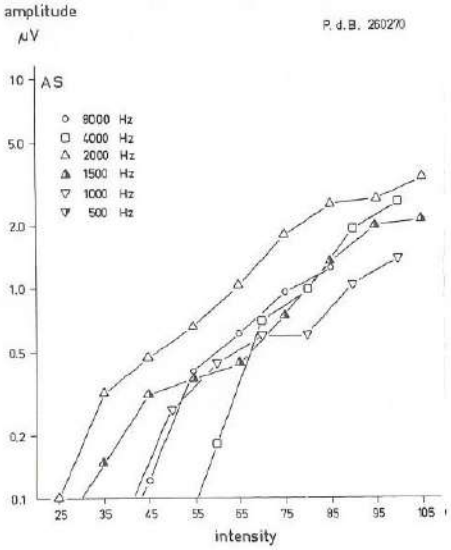
ECoG was performed on both ears. The figures on the opposite pages refer to the results of the left ear.
Both middle-ears contained glue, which was sucked out before the electrocochleogram was made.
In the *left* ear, thresholds were 20 dB from 500 up to 2000 Hz, dropping to 45 dB at 4000 and 8000 Hz (Fig., upper left). Input-output curves were normal for 500, 1000 and 2000 Hz, but showed recruitment for 4000 and 8000 Hz. The amplitude-latency curve was outside the 3σ-boundaries for 8000 Hz but normal for the other frequencies. Summating potentials were of negative polarity for 2 and 4 kHz, having rather small amplitudes: $-0.3\mu V$ for 2000 Hz, 75 dB and $-0.9\mu V$ at 90 dB, 4000 Hz. For 8000 Hz, however, a positive summating potential was re-

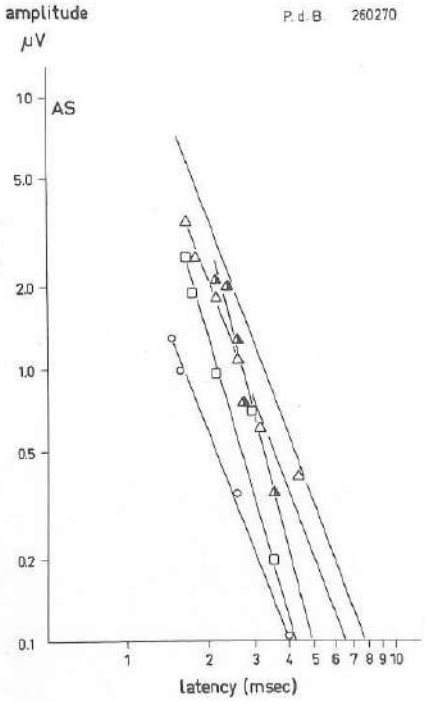
corded: $+0.7\mu V$ at 85 dB. These results indicate a cochlear hearing loss for 4000 and 8000 Hz on basis of the slope of the input-output curves. Outer haircell loss is expected, considering the positive summating potential for 8000 Hz. The threshold elevation for frequencies below 2000 Hz is contributed to a remaining conductive component of 20 dB, due to the tubotympanitis. The results of the electrocochleogram at the *right* ear were approximately similar. Here too a cochlear hearing loss was found for frequencies above 2000 Hz with outer haircell damage for 8000 Hz. The conductive component at this ear was 40 dB.

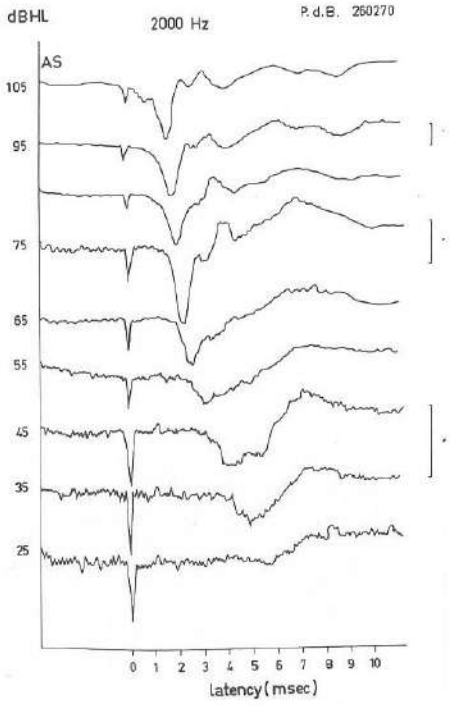
Discussion

Despite the removal of the glue directly before the electrocochleogram was made, a considerable conductive loss was measured. In our experience this is not unusual and is probably due to some residual glue on the ossicular chain and the round window.
Directly after ECoG middle ear ventilation tubes were placed. After a few months it became possible to perform observation audiometry. The results confirmed the electrocochleographic findings of a high-frequency hearing loss, while lower frequencies were now normal. The sensori-neural origin of the hearing loss was already revealed by electrocochleography. This type of the audiogram is frequently found in progressive hearing loss. (Huizing and Carrière, 1975). Therefore, observation audometry is repeated twice a year and until now no progression of the hearing loss has been demonstrated.









A 1.5 Autistic child with mixed hearing loss

Case nr. 260270

History

A 3 year old boy with a history of asphyxia at birth, kernicterus and epilepsy, was sent to our clinic. Although the parents were convinced that his hearing was normal, his pediatrician questioned the child's hearing ability. No understandable speech had been developed yet. It was difficult to get in touch with the child, which showed serious psychological problems and autistic behaviour.

Examinations

Bluish eardrums were observed, but no fluid was found upon paracentesis. The tonsils were large, as was the adenoid.

Audiometry

Although observation audiometry was performed several times, no good reactions could be obtained to standard sounds as the child was totally uncooperative.

Indications for electrocochleography

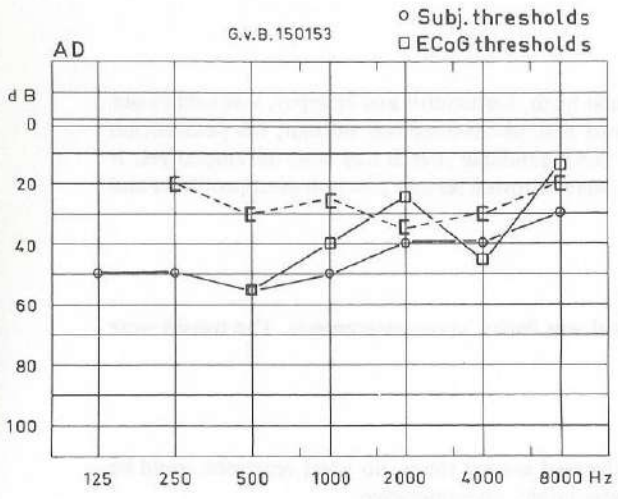
It was of major importance to have exact information about he hearing ability of this child with serious psychological problems.

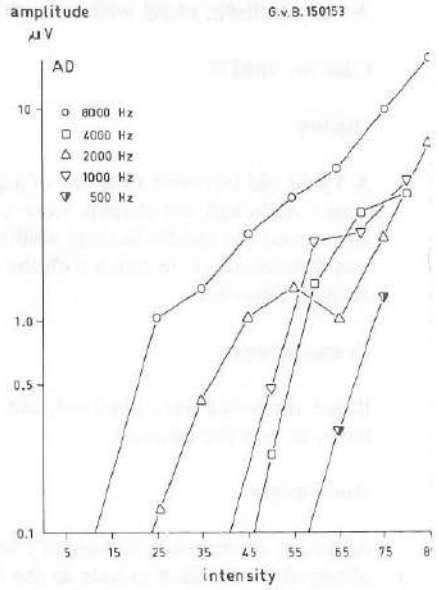
Results of electrocochleography

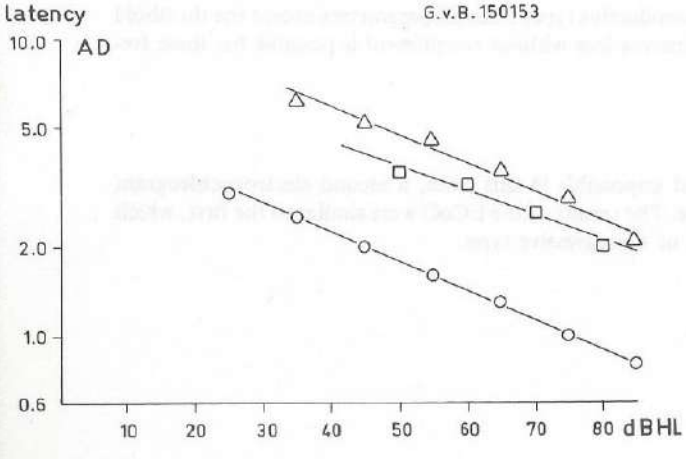
ECoG was performed on the left ear. Thresholds were (Fig., upper left) 45 dB for 500, 40 dB for 1000 and 20 dB for 2000 Hz. For 4000 and 8000 Hz thresholds were 55 and 45 dB respectively. The slope of the input-output curves was normal for all frequencies. The amplitude-latency curve was normal for 2000 Hz, but abnormal (outside 2σ boundaries) for 4000 and 8000 Hz. The SP at 2000 Hz was $-0.6\,\mu V$ at 85 dB. For 4000 and 8000 Hz no SP could be detected, what is indicative for hair-cell loss for these frequencies. For frequencies below 2000 Hz, the hearing loss might be of the conductive type, since all parameters except the threshold are normal. However, any type of hearing loss without recruitment is possible for these frequencies.

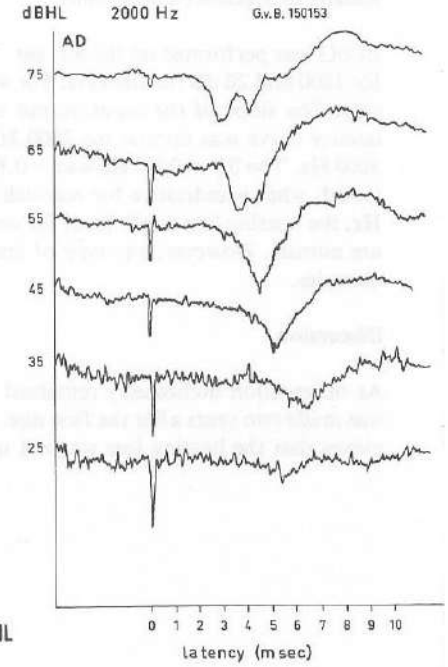
Discussion

As observation audiometry remained impossible in this child, a second electrocochleogram was made two years after the first one. The results of the ECoG were similar to the first, which means that the hearing loss was not of a progressive type.









A 2.6 Suspected aggravation turns out to be otosclerosis

Case nr. 150153

History

A 20 year old male complained of a bilateral slowly progressive hearing loss. However, on the admission-examination for the Dutch army, his hearing was reported to be normal. His family-doctor, knowing that the family history suggested otosclerosis, referred him to our clinic.

Examinations

Eardrums and tubal functioning were normal. Physical examination showed a healthy young man. X-rays of the petrous bones showed normal internal auditory canals.

Audiometry

The first audiogram showed a pure perceptive-type hearing loss of 50 dB at the left ear with a flat curve, and at the right ear a mixed hearing loss of 70 dB with a conductive component of 20 dB. A few days later a 40 dB mixed loss was recorded at the left ear as well as at the right one. Speech audiometry at the left ear showed a maximum discrimination of 100% with a 20 dB shift of the curve to normal. The curve at the right ear showed a 30 dB shift, and here too a 100% discrimination was reached. Sisi-tests invariably showed 0 percent scores. Tone decay tests were normal, except for a dubiously pathological decay for 2000 Hz at the left ear. Békésy audiograms for both ears showed a ± 20 dB greater hearing loss for the pulsating tone than for the continuous one (Type V). Stapedius reflexes were negative for both ears.

Indications for electrocochleography

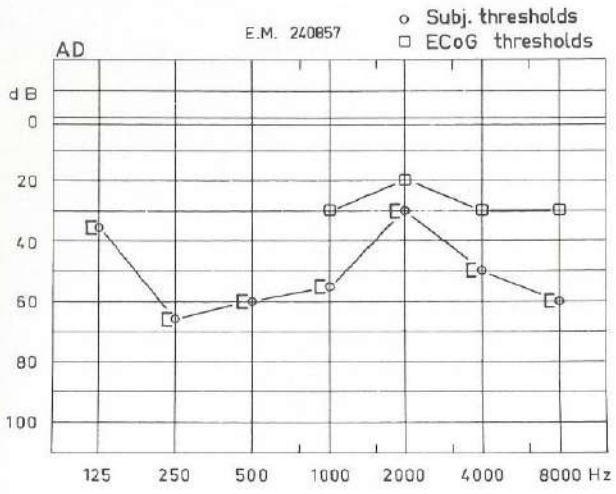
Although a hearing loss was likely, the exact amount and type of the loss could not be determined. The threshold tests were repeated several times, but no consistent results could be obtained. Although simulation was suspected, the cause of the different audiometric results remained obscure.

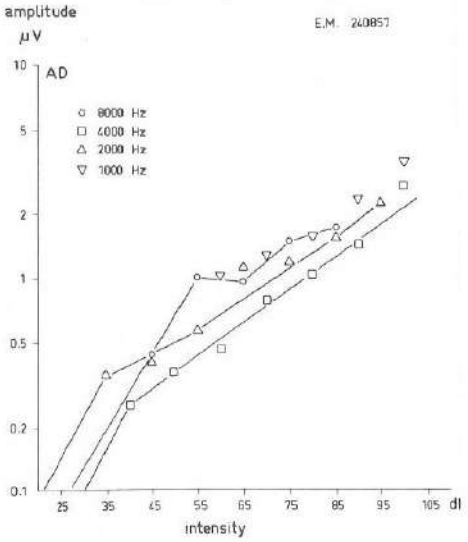
Results of electrocochleography

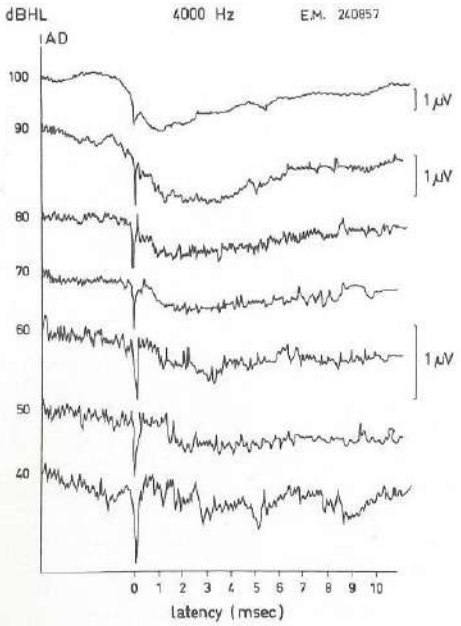
Electrocochleography was performed on the right ear. AP thresholds were in agreement with thresholds found in one of the most recent audiograms (Fig., upper left). For speech frequencies an average loss of 50 dB was found. Input-output curves (Fig., upper right) were normal for 2000 and 8000 Hz, but showed steeper slopes and a smaller maximum output for 500, 1000, and 4000 Hz, than for 8000 Hz. This implies that recruitment is absent for these frequencies. Summating potentials for 2000, 4000 and 8000 Hz were all of negative polarity, amplitudes being small, but within normal range ($-1.2\ \mu V$ for 2000 Hz at 85 dB, $-0.6\ \mu V$ for 4000 Hz at 70 dB and $-2.4\ \mu V$ for 8000 Hz at 85 dB). The normal SP amplitudes exclude a haircell loss. Latency-intensity functions showed a normal curve for 8000 Hz, but a 10-15 dB shift from the average normal curve for 2000 and 4000 Hz, suggesting a 10-15 dB bone-air gap for those frequencies.

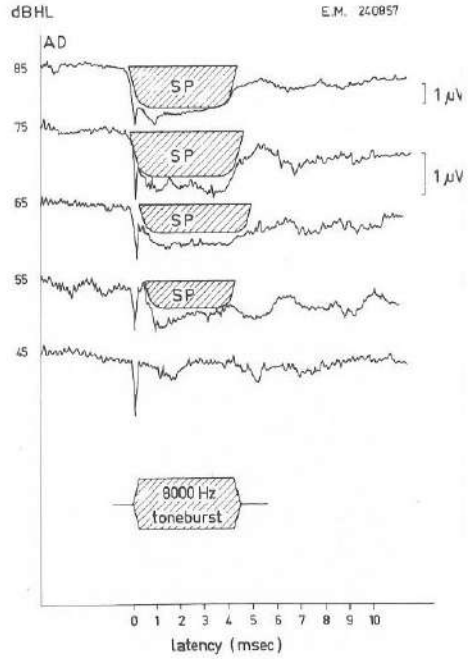
Discussion

The rather confusing results of the different and successive hearing tests suggested the possibility of a non-organic hearing loss in this young man with a family history of otosclerosis. The electrocochleographic data, however, demonstrated a considerable organic hearing loss. This, together with the positive family history, the normal eardrums and the negative stapedius reflexes, made the diagnosis otosclerosis more likely, although no larger bone-air gap could be demonstrated. The middle ear was opened and a totally immobile stapes, due to otosclerosis, was found. Stapedectomy was performed and a gelfoam-wire prosthesis was placed. Two months later the audiogram had improved by 15-25 dB for frequencies below 2000 Hz.









A 2.7 A 'psychogenic' deafness proves to be an organic hearing loss

Case nr. 240857

History

A 16 year old boy complained of a hearing loss and heavy tinnitus in both ears for the past four years. He was examined by several ENT specialists, who found it extremely difficult to obtain a reliable audiogram, and who were inclined to consider the hearing loss as 'non-organic'. He was referred to a psychiatrist who started psychotherapy. It was suggested that the fact that the boy was dominated by his physically stronger twin brother could play an important role and give raise to a simulated hearing loss. Results at school were moderate.
The parents and the boy requested a new ENT and audiometrical examination at our hospital.

Examinations

Normal eardrums and normal tubal functioning were found. Röntgenography of mastoids and petrous bones was completely normal.

Audiometry

Subjective audiometry was performed many times. The boy's reactions were very insecure and uncertain. Now and then, he seemed to be in 'another world'. Nevertheless, after many audiograms, it was believed that the boy's hearing loss was, at least partially, organic. Pure tone audiograms at the right ear showed a 50 dB perceptive loss over the whole frequency range. The left ear also showed a 50 dB perceptive loss, but here the lower frequencies were more affected than the higher.
Speech audiometry, however, invariably showed very poor discrimination. The Békésy audiogram showed a 40 dB loss at the left ear both for continuous and pulsating stimuli. At the right ear, a 50 dB hearing loss was recorded.
However, the curve of the continuous tone was far below the curve of the pulsating tone (Békésy type IV).

Vestibulometry

The electronystagmogram showed a very small spontaneous nystagmus to the left side in all headpositions. For each ear the caloric excitation with water of 33°C caused a nystagmus. On stimulation of the right ear the nystagmus was more pronounced and persisted longer than on stimulation of the left ear. No nystagmus was recorded on excitation of the ears with water of 44°C.

Indications for electrocochleography

Although an organic hearing loss was suspected, proof, however, could not be given by subjective audiometry. Moreover, the Békésy audiogram at the right ear showed a type IV curve, suggesting a retrocochlear hearing loss. It was decided to perform ECoG to determine the exact hearing thresholds and, in case of a hearing loss, to supply more information about the site of the lesion.

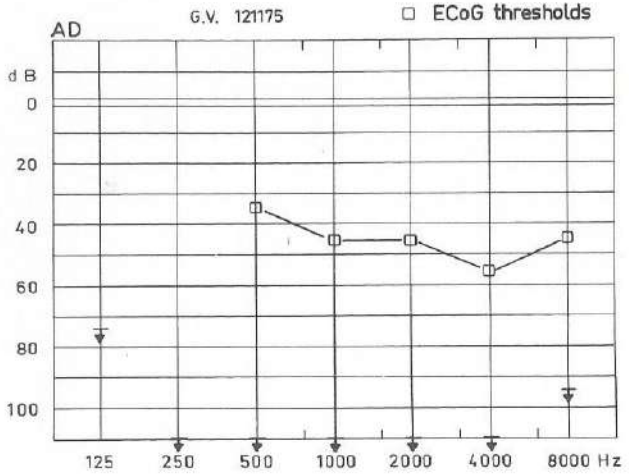
Results of electrocochleography

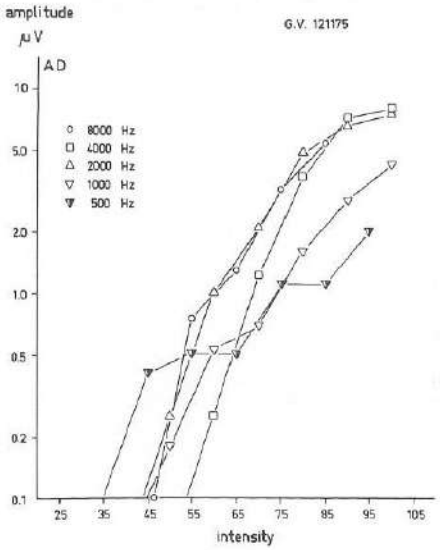
ECoG was performed on both ears. No significant difference was found between both ears. Therefore only the results of the right ear are presented and discussed. Thresholds at the right ear were 25 dB average over the whole frequency range. The slope of the intensity-amplitude curve was abnormally flat (Fig., upper right).
The flatness of the curve is probably caused by desynchronization of the action potentials (Fig., lower left) or by a diffuse loss of auditory nerve fibres. This suggests a disturbance in the functioning of the cochlear nerve. The negative polarity and the normal amplitudes of the SP demonstrate that cochlear function is normal (Fig., lower right). (Note how nicely in this case the trapezoidal shape of the stimulus is represented by the SP).

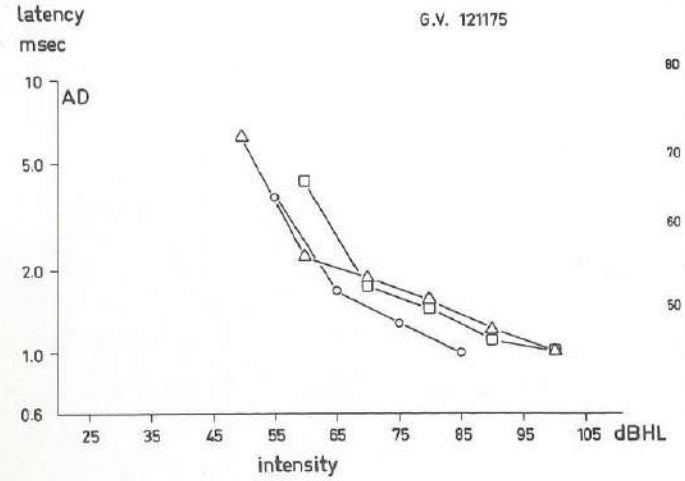
Discussion

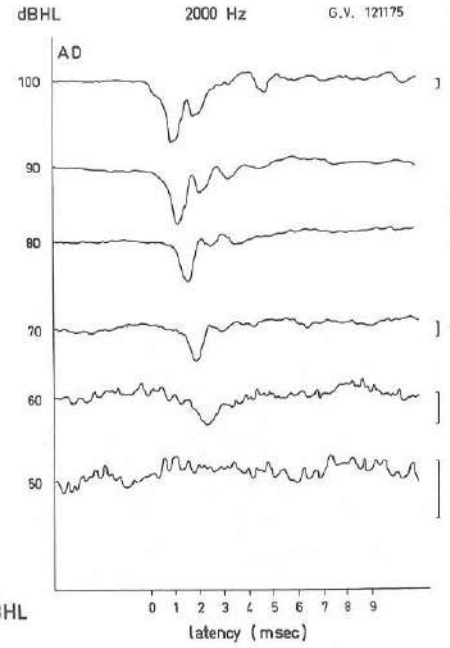
The very poor speech discrimination as related to the good cochlear function could be explained by desynchronization of the action potentials, resulting in a loss of information between the cochlea and the brain.

Since the left ear showed almost exactly the same recordings, a disseminated neural disturbance is likely. Vestibulometry showed a spontaneous nystagmus to the left side and a possible diminished excitability of the left labyrinth.
An electroencephalogram showed diffuse and unspecific disturbances. Further extensive neurological examination did not reveal abnormalities.
The psychiatric treatment of the supposed not organic hearing loss was stopped when the organic nature of the hearing loss was established by electrocochleography.
It should be realized, however, that the exact nature and cause of the hearing loss could not be elucidated.









A 2.8 Aggravation of the hearing loss in the better ear in a case of a one-sided deafness

Case nr. 040414

History

A 61 year old male patient was sent to our clinic because of severe hearing impairment of both ears. The patient had become almost totally deaf after a fracture of the skull base many years ago. The question put to us was whether we could rehabilitate this patient with a powerful hearing aid. The history was very difficult to take. He complained of almost total deafness and heavy tinnitus in both ears. Also slight dizziness was reported to be present continuously.

Examinations

Routine ENT examination showed normal eardrums and good tubal functioning. General physical examination showed an enlarged liver and disturbed liver functions. This was probably due to alcohol abuse. Neurological examination showed no additional features. Tomography of the petrous bones showed bilateral sclerosis, it was not possible to visualize the left cochlea, but the right cochlea had a normal appearance. The internal auditory canal on both sides was normal.

Audiometry

The pure tone-audiogram showed no responses at both ears. Impedance measurements showed normal patterns in both ears. The stapedius reflex was negative in the left ear, but there were slight reactions in the right ear on stimulation with 100-110 dB. In speech-audiometry, no discrimination was found.

Vestibulometry

Electronystagmography showed a slight nystagmus to the right in the left-side position. The left labyrinth was unexcitable on caloric stimulation.

Indications for electrocochleography

Although pure tone-audiometry showed a 100-110 dB loss at the right ear, the stapedius-reflex test suggested a smaller loss.
ECoG was indicated, firstly to record the exact hearing thresholds, secondly to reveal the possible retrocochlear component in the hearing loss.

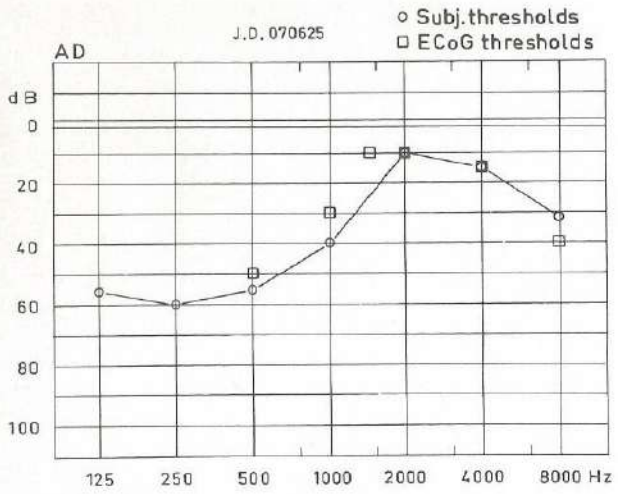
Results of electrocochleography

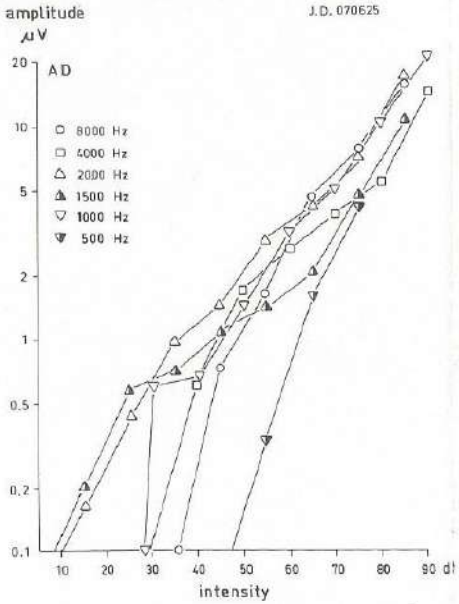
Electrocochleography was performed on the right ear. Thresholds were far better than those found in subjective audiometry, now a 45 dB average loss was found. Recruitment was present for 2000, 4000 and 8000 Hz (Fig., upper right). The latency-intensity curves were within the normal range, although some resemblance could be seen with curves found in retrocochlear processes (Fig., lower left). Summating potentials were all of negative polarity, having small amplitudes: 2000 Hz, 80 dB $-0.8\,\mu V$, 4000 Hz, 80 dB $-0.2\,\mu V$, 8000 Hz, 85 dB $-0.9\,\mu V$. It was concluded that the hearing loss was of cochlear origin, showing recruitment. To a limited extent outer haircell lesions in the basal turn were likely.

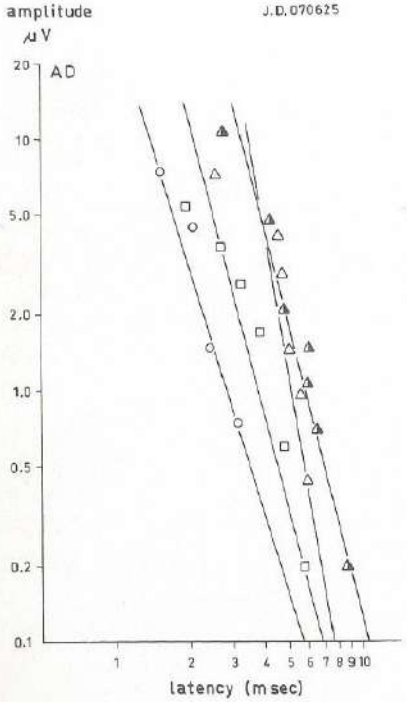
Discussion

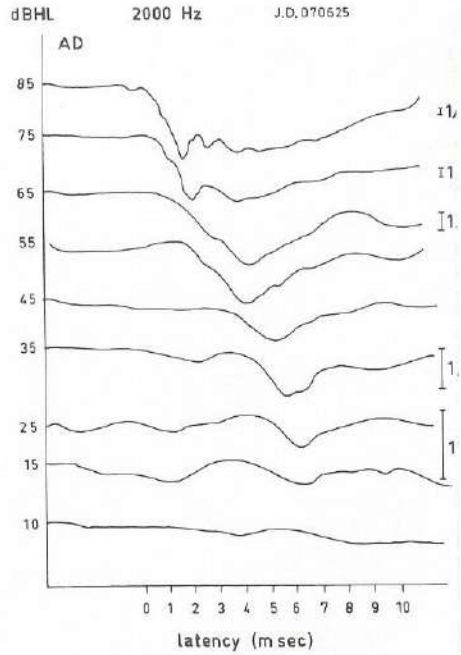
The ECoG results were discussed with the patient, and a new pure tone audiogram was made. Now the left ear still showed a total hearing loss, but the right one only a 65 dB average loss in speech frequencies.

As a result of the electrocochleographic diagnosis, it became evident that our patient aggravated, although he was a hard of hearing patient, handicapped by his disability. A hearing aid was fitted, with which our patient was fairly well helped. It remained obscure why this patient pretended an almost complete hearing loss on both sides initially.









B 1.9 A classical case of Menière's disease

Case nr. 070625

History

A 51 year old male presented himself at our clinic complaining of recurrent attacks of vertigo, unilateral tinnitus and hearing loss, mostly during periods of stress, which were accompanied by vomiting. During an attack his hearing became worse in the affected right ear.

Examinations

The aspect of the eardrums was completely normal and so was tubal functioning. At general physical and neurological examination no abnormalities were found. X-rays made of the petrous bones and the cervical spine were normal, and clinical blood chemistry showed normal values.

Audiometry

During hospitalization audiometry was performed several times. The left ear showed normal hearing, but at the right ear a perceptive loss at frequencies below 2000 Hz was found up to 60 dB at 250 Hz. Right-ear speech audiogram curves were 10-30 dB shifted with respect to normal, with a maximum discrimination score of 90%. Although Sisi scores were invariably low, ABLB tests showed recruitment for 500 and 1000 Hz. Tone decay tests were normal. The Békésy audiograms, both for continuous and interrupted stimuli, were in good agreement with the thresholds found by pure-tone audiometry.

Vestibulometry

No spontaneous nystagmus was recorded, and both labyrinths were equally excitable with cold and warm water.

Indications for electrocochleography

Conventional audiometric findings suggested a perceptive hearing loss with recruitment. To establish its cochlear origin, and to exclude retrocochlear involvement, and also to reveal the possible presence of (outer) haircell lesions, ECoG was indicated.

Results of electrocochleography

AP thresholds for the right-ear agreed with thresholds found in conventional audiometry. Input-output curves showed recruitment for 500, 1000, 4000 and 8000 Hz (Fig., upper right), amplitude-latency curves were within normal range for the recorded frequencies (1500, 2000, 4000 and 8000 Hz). Summating potentials were constantly of negative polarity and amplitudes were very large (ranging from $-2.1\ \mu V$ at 4000 Hz to $-8.4\ \mu V$ at 2000 Hz). The hearing loss was concluded to be of pure cochlear origin, being of the metabolic type, as it showed no outer haircell loss, except for 8000 Hz, for which frequency no SP could be detected.

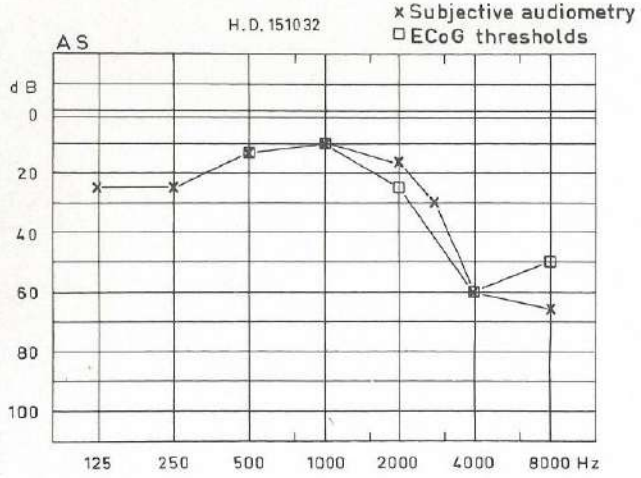
Discussion

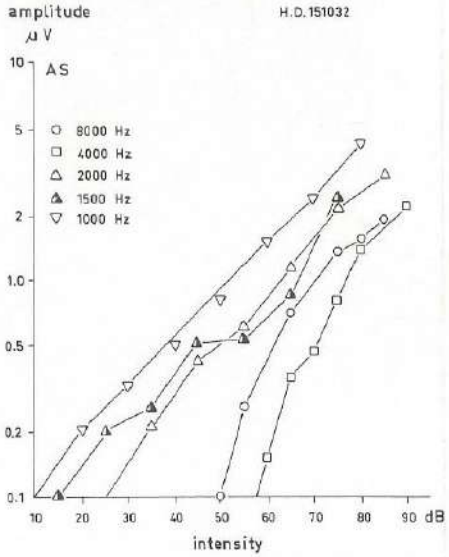
The large – at least larger than normal – negative summating potential is a phenomenon

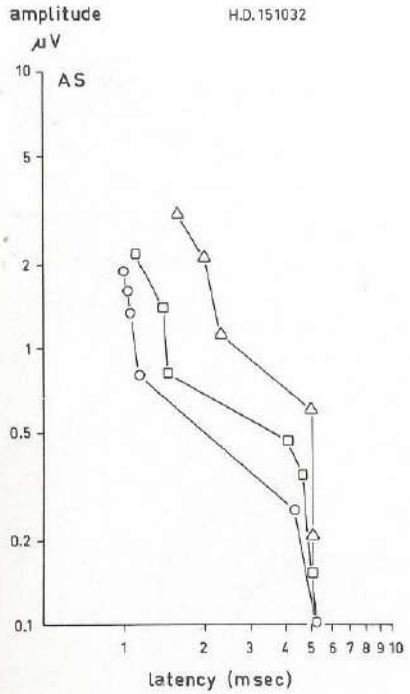
which, in our experiences, is only observed in patients suffering from Menière's disease. This may be due either to metabolic dysfunctioning or to a static displacement of the basilar membrane towards the scala tympani, thus reflecting the presence of a hydrops. Since summating potentials were normal at 2 and 4 kHz, the hearing loss should (theoretically) be reversible at these frequencies.

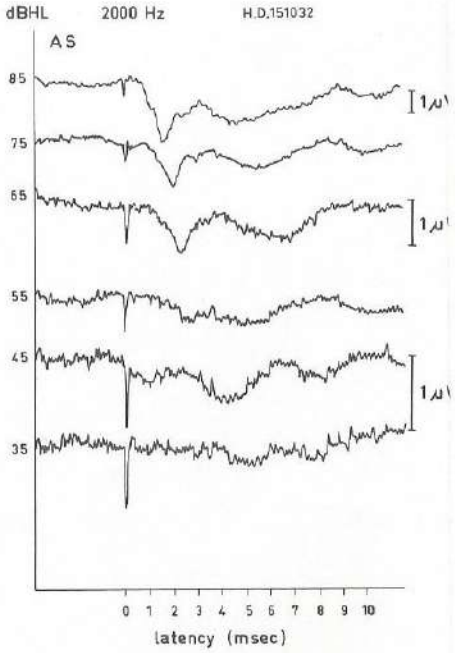
The diagnosis of Ménière's disease was made on basis of history, conventional audiometry and electrocochleography. The absence of a spontaneous or positional nystagmus in vestibulometry is not in contradiction with this diagnosis, although a difference in the slow-phase velocity of the nystagmus due to caloric stimulation between the normal and the pathological ear is found in 80% of the patients with Menière's disease in the interval between the attacks of vertigo (Baarsma, 1979).

Treatment consisted of a vasodilator regimen, in this case intravenously administered histamine. Sedation and cinnarizine were also given. Now, two years later, vertiginous attacks are less frequent, although hearing and tinnitus have not improved.









B 1.10 Fluctuating hearing loss in one-sided deafness

(Cochlear stage of Menière's disease)

Case nr. 151032

History

A 42 year old male patient was known to us because of a previous operation for chronic middle ear disease of his right ear. He was totally deaf in this ear since many years. Two months after the operation our patient visited the hospital complaining of serious hearing impairment in his left ear. He told us that this happened at least three times a year, especially when he suffered from a cold.
After a few days, hearing always had returned to normal. While his hearing was impaired, he also noticed a slight dizziness. We decided on a clinical observation.

Examinations

On the left side a normal eardrum and normal tubal functioning was found. The right ear showed a tympanic membrane as seen after tympanoplasty. The mucous membranes of the nose showed a slight redness as is usually seen in colds. X-rays of the petrous bones showed no abnormalities. General physical and neurological examination showed no abnormalities. Blood chemistry values were normal. The spinal fluid showed a slightly elevated protein content.

Audiometry

The right ear showed a total hearing loss. Thresholds at the left ear were 65 dB perceptive loss in a flat curve. The same day the audiogram of the left ear improved by 15 dB to a 50 dB perceptive loss, flat curve. Two days later, thresholds at speech frequencies were 15 dB, at lower frequencies 35 dB and 50 dB for 4000 and 8000 Hz, all being a pure perceptive loss. Another two days later, the patient told us that his hearing was poor again, and that, before hearing became worse, he had noticed tinnitus in his left ear. The audiogram showed again a 60 dB flat perceptive loss.

Vestibulometry

Positional tests showed a deviation to the left side. Nystagmography showed a spontaneous nystagmus to the left side. The left labyrinth was significantly less excitable on caloric stimulation.

Indications for electrocochleography

Although Menière's disease was suspected, the typical vertiginous attacks were absent. The possibility of multiple sclerosis was also considered on basis of the fluctuating hearing loss, the slightly increased protein content of the spinal fluid, and the spontaneous nystagmus. ECoG was performed to differentiate between those two possibilities.

Results of electrocochleography

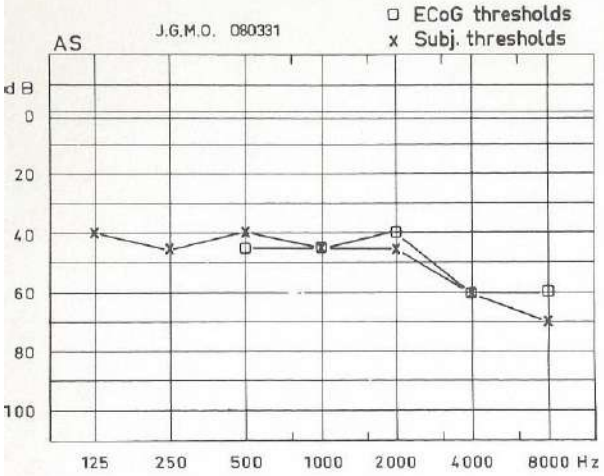
ECoG was performed the day after the audiogram for the speech frequencies showed only a small perceptive loss.
Thresholds corresponded well with thresholds found in subjective audiometry the day before.

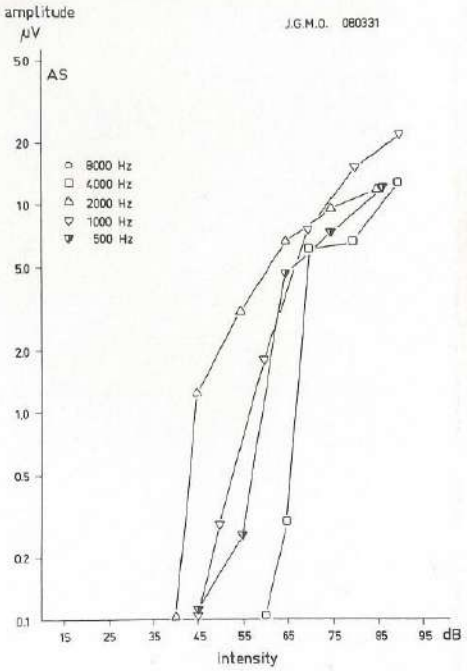
Input-output curves showed a steep slope for 4000 and 8000 Hz, which meant that recruitment was present for these frequencies. For the other frequencies the normal slope of the input-output curves indicated the absence of recruitment. Amplitude-latency curves were abnormal for 4000 and 8000 Hz. The absence of summating potentials for 4 & 8 kHz suggested outer haircell loss. Summating potentials for 2000 Hz were of negative polarity and had normal amplitudes ($-1.5\ \mu V$ at 85 dB).

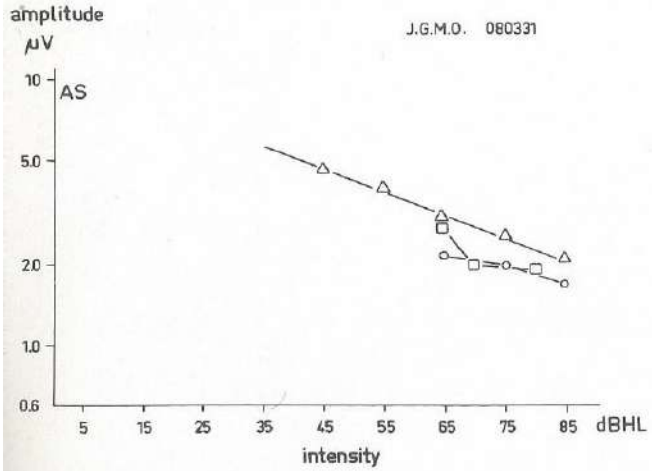
A few months later a second electrocochleogram was made. Our patient still suffered from attacks of hearing loss. This time, the electrocochleogram was made the day after the audiogram showed a 70 dB perceptive loss. ECoG thresholds again agreed well with thresholds found in subjective audiometry. Now input-output curves showed recruitment for 2000, 4000 and 8000 Hz. Amplitude-latency curves were abnormal for 2000, 4000 and 8000 Hz. Summating potentials were absent for 8000 Hz, very small ($-0.3\ \mu V$ at 80 dB) for 4000 Hz and normal ($-1.2\ \mu V$ at 85dB) for 2000 Hz. Consequently, outer haircell lesions seemed likely for 4000 and 8000 Hz, which confirmed the data obtained in the first electrocochleogram. Recruitment was also present for 2000 Hz, which was not observed in the first ECoG. Narrow-band responses for 2000 Hz showed the triphasic waveform, which is typical for Menière's disease. It was concluded that the results of the second ECoG pointed to a metabolic disturbance of the cochlea (with outer haircell lesions for 4000 and 8000 Hz), which very well could fit with Menière's disease.

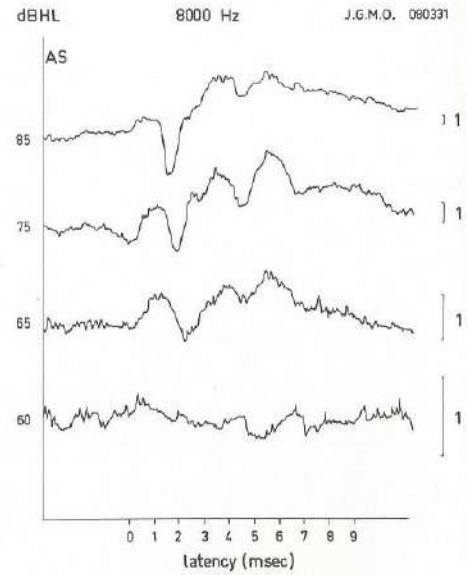
Discussion

ECoG excluded a retrocochlear lesion as could have been expected in multiple sclerosis, and confirmed a hearing loss of cochlear origin with outerhaircell lesions, thus indicating a metabolic disturbance as is found in Menière's disease. Although the typical vertiginous attacks are absent, it is expected, that in due time the complete syndrome of Menière will develop. This could be called the cochlear stage of Menière's disease (Schmidt et al. 1979).









B 1.11 Bilateral Menière's disease?

Case nr. 080331

History

A 42 year old business-man was referred to our department by the neurologist of another hospital. He complained of fluctuating hearing loss in both ears, present for the past year. Tinnitus was absent. Sometimes a slight vertigo of short duration had been present. The neurologist reported no abnormalities on physical examination, especially no cerebellar symptoms had been found. However, on angiography of the carotid arteries a stop in the right medial cerebal artery had been found. As this unexpected finding could not explain the patients complaints, he was referred to our department.

Examinations

Routine ENT examination showed no abnormalities. X-rays of the petrous bones showed a normal diameter of both inner ear canals.

Audiometry

Pure tone audiometry was repeated several times, demonstrating a fluctuating hearing loss between 50 and 90 dB in both ears. The Fig., upper left refers to the left ear audiogram the day before electrocochleography was performed. Speech audiometry showed a loss of discrimination at high intensities. Sisi-scores were invariably high for all frequencies. The tone decay was faster than normal just above threshold for 4000 Hz, but normal at 20 dB above threshold.

Vestibulometry

No nystagmus was seen in electronystagmography; however, on caloric stimulation both labyrinths were less excitable than normal.

Indications for electrocochleography

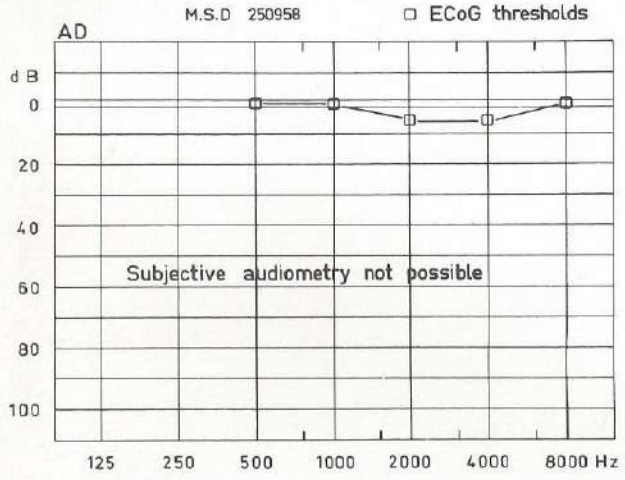
Although the audiometric result suggest a cochlear lesion, the history certainly is not typical for Menière's disease. Furthermore, the angiographically detected stop in the medial cerebral artery suggests a lesion of the central nervous system.
In order to differentiate between a peripheral and a central lesion, it was decided to perform electrocochleography.

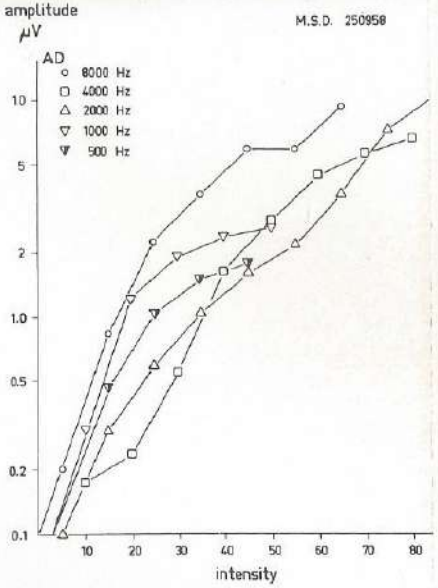
Results of electrocochleography

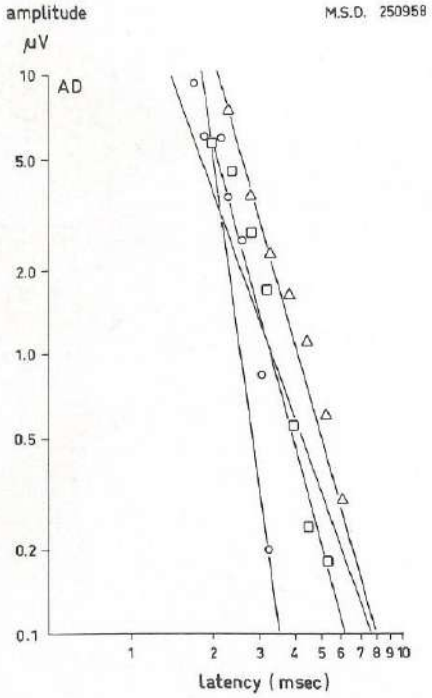
Electrocochleography was performed on the left ear. Thresholds agreed well with those found the day before in subjective audiometry. This suggests a peripheral origin. The slope of the input-output curve was steep for all recorded frequencies, indicating recruitment. The latency-intensity curve was within normal range for 2000, 4000 and 8000 Hz. Summating potentials were of negative polarity for 2000 and 4000 Hz, having normal amplitudes (2000 Hz 85 dB, $-3.5\ \mu V$, 4000 Hz 80 dB, $-2.8\ \mu V$). For 8000 Hz, 85 aB a positive summating potential of $+1.2\ \mu V$ was recorded, indicating outer haircell damage in the most basal part of the cochlea.

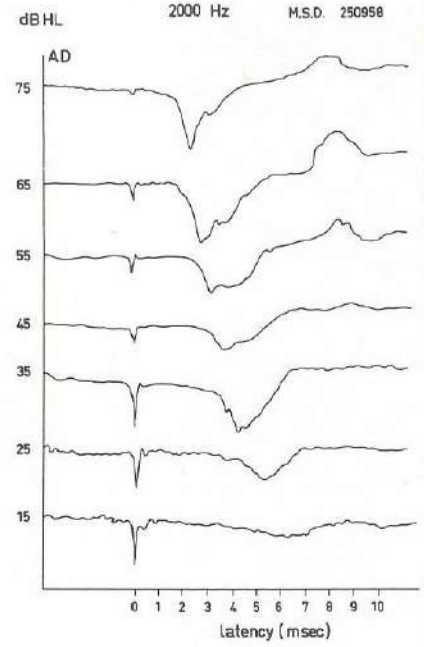
Discussion

When the electrocochleographic thresholds are the same as those found in subjective audiometry, the hearing loss is peripheral. All other electrocochleographic data point to a metabolic hearing loss, as is found in Menière's disease. Therefore the diagnose Menière's disease is likely, but a few uncommon facts must be kept in mind: the patients history is not typical for Menière's disease, while a bilateral fluctuating hearing loss, which is completely symmetrical, is unusual. Finally, the stop in the medial cerebal artery remains unexplained.









B 2.12 Axial ponstumor without impairment of the auditory periphery

Case nr. 250958

History

A seventeen year old girl was admitted in the department of neurosurgery. She had been suffering from progressive symptoms of increased intracranial pressure, i.e. bi-frontal headache, serious vomiting in morning hours, and blurred vision. The neurologist found a bilateral paresis of the third, fourth, fifth, sixth and nineth nerve, the right side being more affected than the left. Due to the patients subcomatous condition, the function of the facial, auditory and vestibular nerves could not be examined. Pneumo-encephalography and angiography showed a large axial tumor of the pons, extending in the left pontine angle.

Examinations

Examinations showed no additional features. Normal eardrums were found. No impression of auditory function could be obtained with subjective audiometry. Impedance measurements were not performed.

Indications for electrocochleography

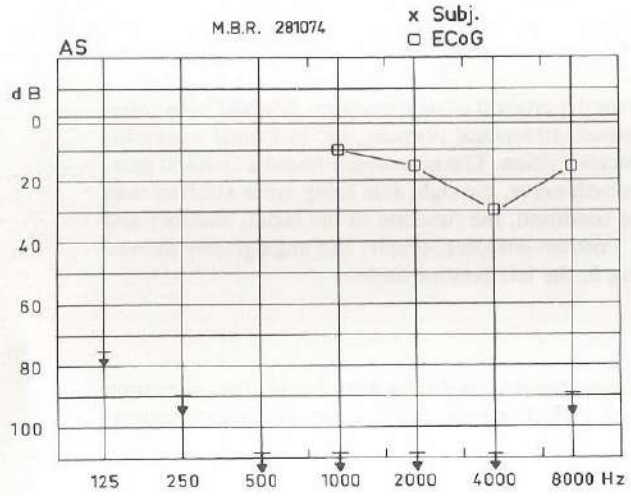
In this case of a suspected axial ponstumor, it was important to know whether the function of the cochlear nerves was affected or not. Impairment of the functioning of the cochlear nerves would indicate invasion of the pontine angles by the tumor, which would make a surgical approach to the tumor much less favorable.
Since subjective audiometry, due to the patients subcomatous condition, was impossible, ECoG was indicated.

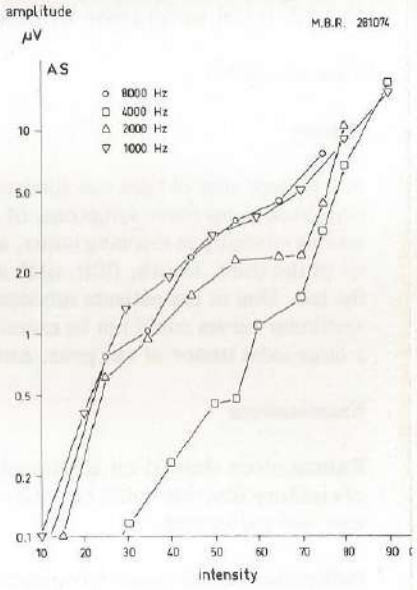
Results of electrocochleography

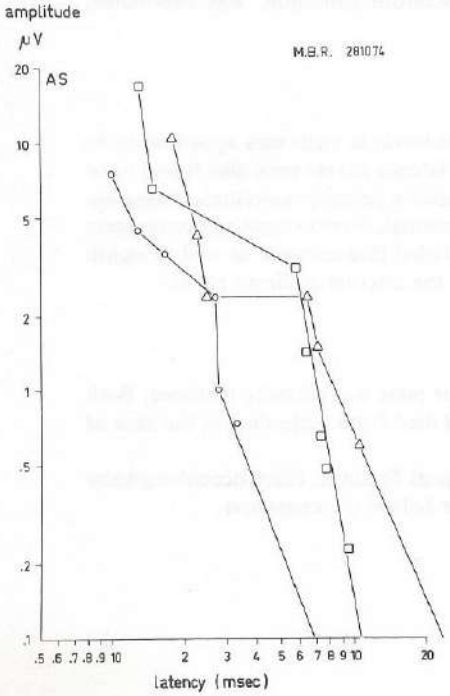
Electrocochleography was performed on both ears. Thresholds in both ears appeared to be within normal limits. Input-output curves and amplitude-latency curves were also found to be normal in both ears. Summating potentials were all of negative polarity, amplitudes being approximately $-1.8\ \mu V$ for 8000 Hz at 85 dB, which was normal. Narrowband responses were diphasic, as is usually found in normal ears. It was concluded that cochlear as well as eighth nerve functioning was completely normal at the level of the internal auditory canal.

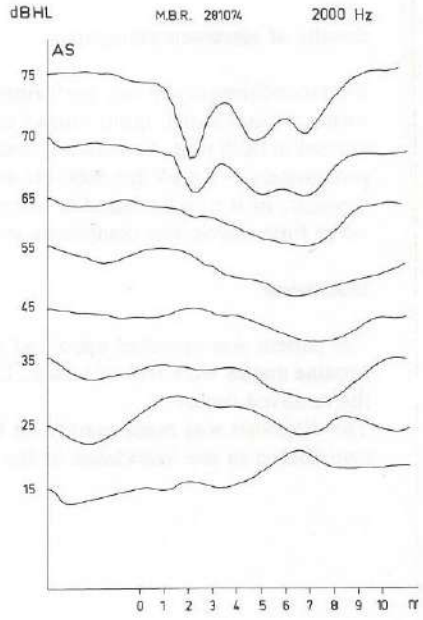
Discussion

The patient was operated upon and an astrocytoma of the pons was partially removed. Both pontine angles were free of tumor. Unfortunately patient died from a bleeding in the area of the removed tumor.
The diagnosis was made mainly on basis of the radiological findings. Electrocochleography contributed to the knowledge of the spread of the tumor before the operation.









B 2.13 A pontine angle neuroma without impairment of the auditory periphery

Case nr. 281074

History

A 54 year old male complained of impaired hearing and of imbalance, causing him to fall to his left side, when riding his bicycle. The patient consulted a neurologist who found papilledema, and an ENT specialist who found an unexcitability of the left labyrinth and total deafness of the left ear. Petrous bone X-rays showed a widening of the inner auditory canal. The patient was sent to our hospital for further evaluation, on the suspicion of a left pontine angle tumor.

Examinations

Ear drums were normal as was tubal functioning. No evidence of damage to the cranial nerves could be found, except for the cochlear and vestibular nerves. Further physical and neurological examination showed no abnormalities, except for the already mentioned bilateral papilledema. On arteriography and ventriculography an axial pons tumor was suspected, but it was not clear if the tumor invaded the left pontine angle. Blood chemistry values were normal. Cerobrospinal fluid showed an elevated protein content.

Audiometry

The right ear showed normal thresholds in pure tone audiometry, but at the left ear responses were absent even at 110 dB HL.

Vestibulometry

Postitional tests showed a deviation to the left side. The left labyrinth was unexcitable upon caloric stimulation.

Indications for electrocochleography

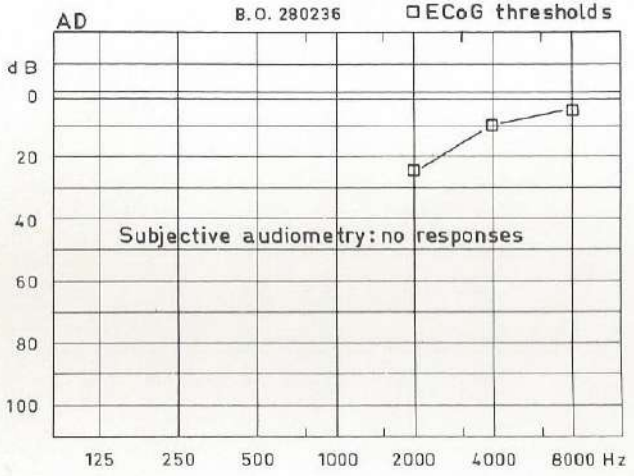
As a total hearing loss was found, it was not possible to determine the locus of the lesion with conventional audiometry. ECoG was used to differentiate between a cochlear and a retrochlear lesion. Furthermore, in case of a retrocochlear one, to predict the spread of the tumor in the left pontine angle, as the latter could not be definitely established by X-ray examination.

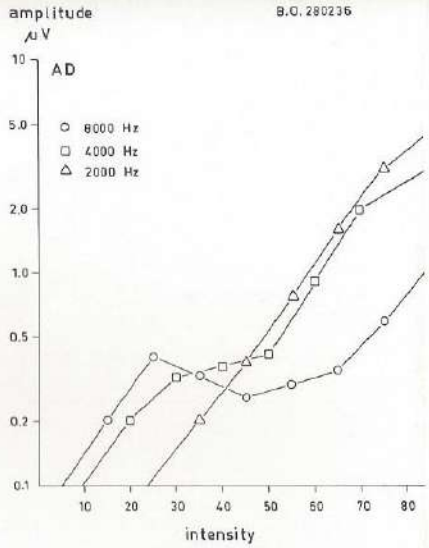
Results of electrocochleography

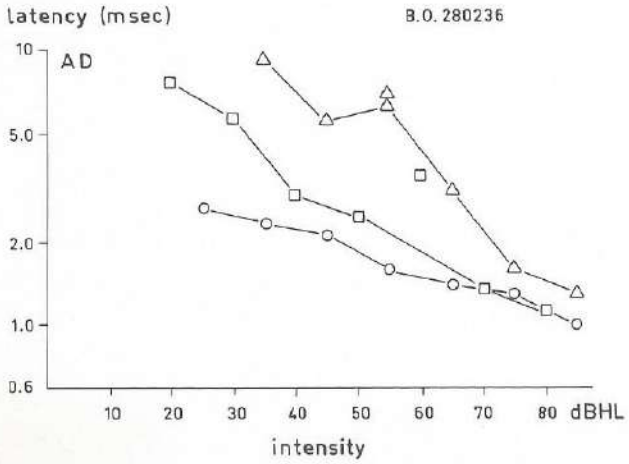
ECoG thresholds at the left ear showed a 10-15 dB loss at speech frequencies and a dip of 30 dB at 4000 Hz, while in subjective audiometry no responses could be obtained. Recruitment was absent for all frequencies, since input-output functions had normal slopes. All amplitude-latency curves were abnormal, passing the upper 2σ – boundary for the normal group, what suggests neural involvement. Summating potentials had normal amplitudes, i.e. $-0.9\ \mu V$ at 75 dB at 8000 Hz and $-3.0\ \mu V$ at 90 dB at 4000 Hz.
It is important to notice that the waveform showed the characteristics usually seen in retrocochlear deafness. The compound action potential consisted of three negative peaks. By lowering the intensity, the N_1 disapeared, while the second and third deflections remained.

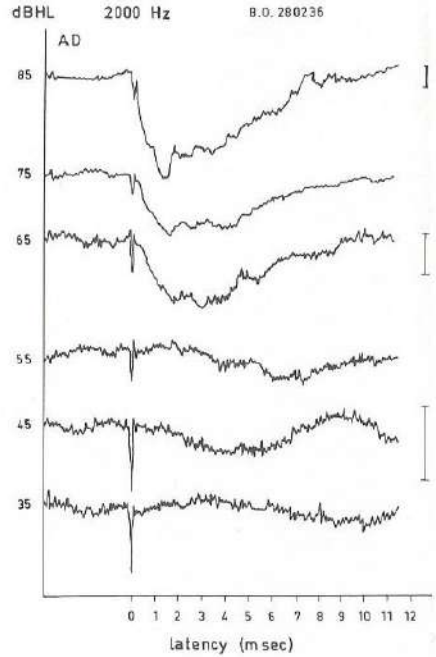
Discussion

The characteristic potential strongly suggested a tumor located near the pons.
If the tumor had invaded the internal auditory canal, diminished blood supply to and from the cochlea would have induced cochlear damage. Since cochlear function was normal, this was not likely to have been the case.
The tumor was explored by the neurosurgeon via a posterior fossa approach. A large tumor compressing the medulla oblongata was removed. The tumor did not invade the bony canal, although it originated from the eight nerve.
All other nearby cranial nerves (VI, VII, IX and X) could be kept undamaged. Histological diagnosis was a neuroma. The suspicion of a pontine angle tumor was mainly established on basis of the history and the neurological and radiological findings, and sustained by electrocochleography. On top of this, electrocochleography contributed to the knowlegde of the spread of the tumor.









B 2.14 Central hearing loss due to brain contusion

Case nr. 280236

History

A 39 year old, previously healthy male patient was hospitalized at the department of neurosurgery, shortly after he had fallen from a roof. Diagnosis of heavy cerebral contusion and cranial fractures of the right temporal bone was made. A month later the patient developed papilledema in the right eye. Tuning-fork tests showed lateralization to the right side. Angiography of the carotid and the vertebral arteries showed a normal pattern of the central vessels. Pneumencephalography did not show abnormalities. The otolaryngologist was consulted with respect to the lateralization test.

Examinations

Routine ENT examination (which was done a month after the accident) showed no anatomical abnormalities. At the tuning fork test this time our patient lateralized to his left ear. Tomography of the petrous bones showed a small longitudinal fracture line in the left petrous bone.

Audiometry

The left ear showed normal hearing thresholds. At stimulation of the right ear no responses were obtained. Tympanometry showed good middle ear pressure.

Vestibulometry

Romberg and Unterberger tests showed a deviation to the right side. No spontaneous nystagmus was recorded and both labyrinths were equally excitable on caloric stimulation.

Indications for electrocochleography

With subjective audiometry a totally deaf right ear was found with a normal functioning labyrinth. No signs of a fracture through the cochlea were seen. No cause had been found for the right sided papilledema. Therefore, to differentiate between a cochlear or retrocochlear hearing loss, ECoG was indicated. In case of a retrocochlear cause, ECoG might be helpful to establish the neurological diagnosis.

Results of electrocochleography

Electrocochleography was performed on the right ear. Thresholds were 25 dB for 2000 Hz, 10 dB for 4000 Hz and 5 dB for 8000 Hz. The slope of the input-output curves was very flat (Fig., upper right). The amplitudes of the summating potentials were normal ($-0.3\ \mu V$ at 2000 Hz, 85 dB, $-2.5\ \mu V$ at 4000 Hz, 80 dB). For 8000 Hz a positive summating potential was recorded ($+1.5\ \mu V$ at 85 dB). The latency-intensity function showed relatively long latencies for 2000 and 4000 Hz (Fig., left below), at intensities below 50 dB.

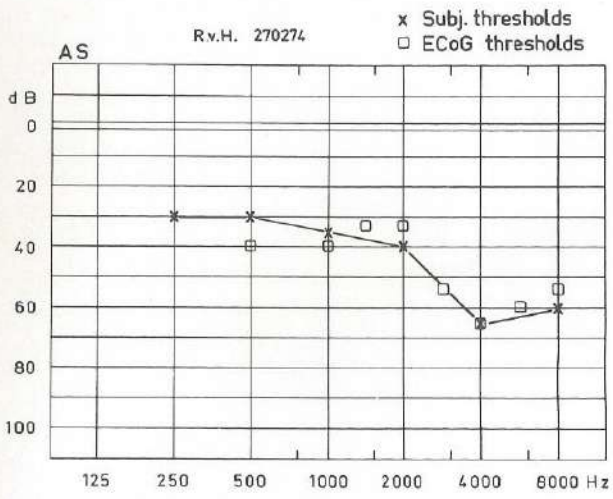
Discussion

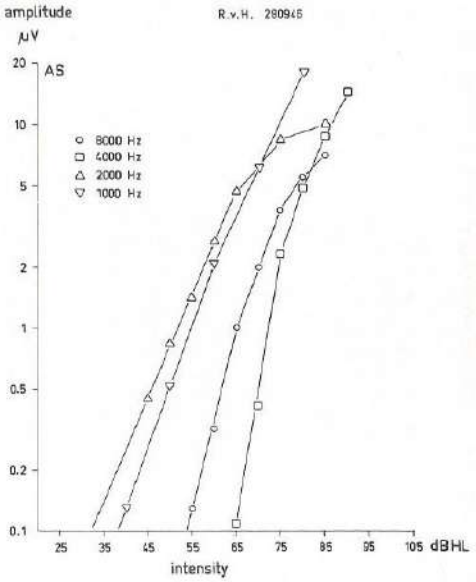
The normal amplitudes of the summating potentials indicated that hair-cell damage was not likely except for 8000 Hz.

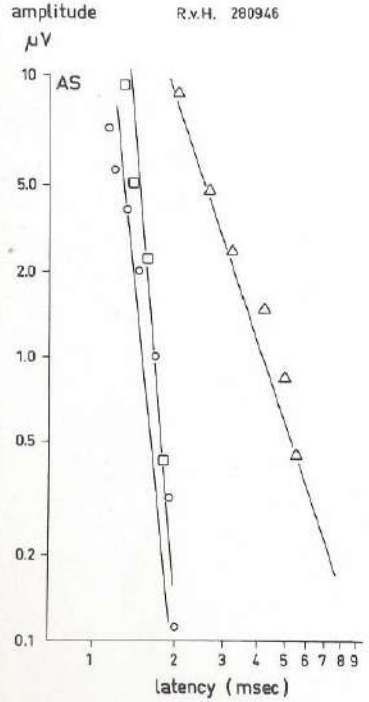
This, and the large threshold-difference between electrocochleography and subjective audiometry, indicated a central cause of the hearing loss, for example a bleeding, or, in this particular case less likely, a tumor.

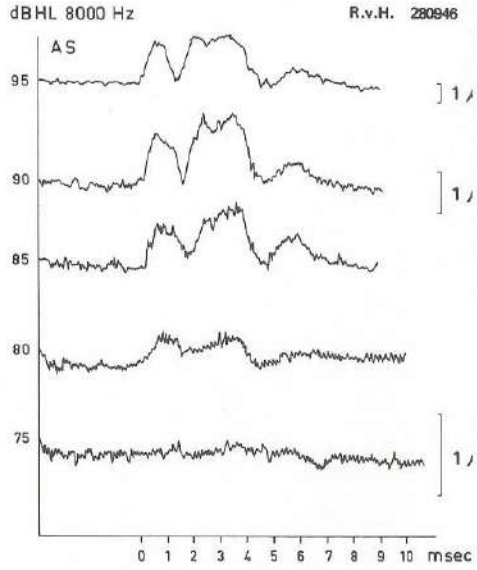
To exclude the possibility of a tumor in the area of the pons cerebri, ventriculography was performed. No abnormalities were seen. The patient's general condition improved spontaneously, papilledema disappeared, but the subjective hearing loss at the right auditory system persisted, for a yet unknown reason. The final diagnosis was: contusio cerebri with contusion of the brainstem.

It should be noted, that if the initial localization of the tuning-fork test to the affected ear was correct, this can only be explained by a conduction-type deafness (as could be expected by a temporal bone fracture), together with a normal to subnormal functioning cochlea and central nervous pathways. This would mean that the central hearing loss developed after the tuning-fork test was done.









B 3.15 Sudden deafness due to syphilis

Case nr. 280946

History

A 28 year old male, complaining of a slowly progressive hearing loss, bilateral tinnitus and attacks of vertigo was sent to our clinic. The hearing loss started 3 to 4 weeks before admittance and the patient complained about aphtaelike eruptions of the oral mucosa one week before the start of his hearing problem, one month before the received an influenza vaccination.

Examinations

Mucosal lesions on the palate and gingivae were found. Ear drums and tubal functioning were normal. The glans penis showed mucosal lesions from which treponema pallidum could be isolated. There were no focal neurological abnormalities, and also X-ray examination of the petrous bone was normal.
The Wasserman and Reiter tests were positive. The spinal fluid showed an increase in cells (68/3) of which 65% were mononuclear. The Queckenstedt test was normal.

Audiometry

A perceptive hearing loss of about 40-45 dB was found bilaterally for frequencies up to 2000 Hz, the higher frequencies showing a 60 dB hearing loss. Tone decay tests and Békésy audiometry did not show any additional features. Sisi scores were 100% for 2000 and 4000 Hz in both ears. Speech audiometry showed a shift of the curves of about 45 dB and a maximum discrimination score of 90%.

Vestibulometry

Positional tests showed no abnormalities. A spontaneous nystagmus to the left side was found. Both labyrinths were equally excitable on caloric stimulation.

Indications for electrocochleography

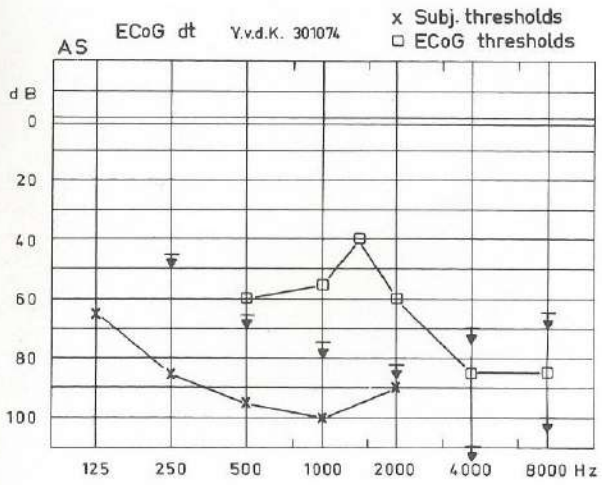
As a syphilitic cause of hearing loss is a rare finding today, the hearing loss was studied more closely by electrocochleography.

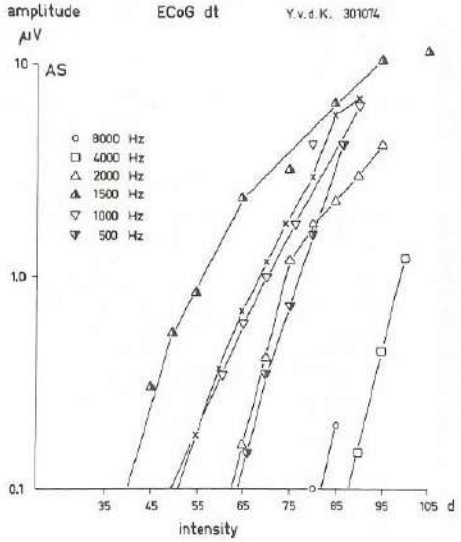
Results of electrocochleography

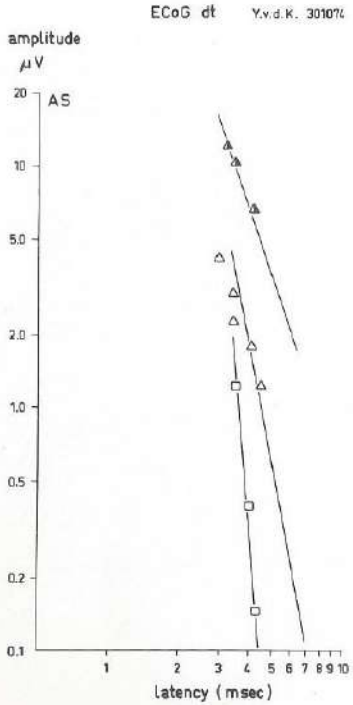
Thresholds agreed well with the subjective ones (Fig., upper left). The input-output curves showed recruitment for 4, 6 and 8 kHz and indications for recruitment at the lower frequencies (Fig., upper right). Amplitude-latency relations were normal for 2 kHz and were outside the normal range for 4 and 8 kHz (Fig., lower left). Summating potentials showed a particular behaviour, being positive at all levels for 8000 Hz, and changing from positive to negative for the lower frequencies with a decrease in intensity (Fig., lower right). The amplitudes ranged up to 7.5 μV for 8000 Hz, 85 dB HL, which is large compared to normal limits and in the range found for Menière's disease. Since normal summating potential amplitudes require normal haircells, but positive SP's indicate at least a loss of haircells in the most basal part of the cochlea, interpretation was not completely clear. It was concluded that the hearing loss would be of the metabolic type which at least for frequencies of 2000 Hz and lower could be reversible.

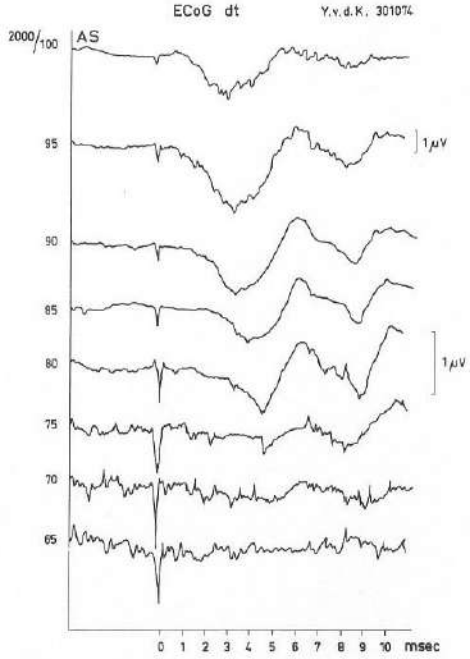
Discussion

A diagnosis of syphilis of stage II has been made. In these cases progressive perceptive hearing loss is known as an infrequent symptom and the hearing loss in general is considered to be of neural origin. Treatment consisted of a daily intramuscular injection of 60.000 IE almocillin during 15 days and appeared to be succesful. No improvement in hearing was seen in the first two months after the treatment was started. However, three months later the audiogram showed normal threshold levels for both ears.
The normal SP's for 2000 and 4000 Hz contradict any, even functional, haircell loss. The positive SP for 8000 Hz suggests, at least a functional haircell loss in the area of the basal turn. The cochleographic data are in good agreement with those found in a large group of Menière patients. The cochlear findings in post mortem temporal bone studies from patients suffering syphilitic hearing loss and those from Menière patients also show no essential difference.
The normal functioning of the haircells indicates a metabolic disturbance which is theoretically reversible.









B 3.16 Sudden deafness of partially central origin

Case nr. 301074

History

An 18 year old girl complained of tinnitus in the left ear and of vertigo, following an attack of a common cold. She consulted an ENT specialist who found a perceptive type hearing loss up to 80 dB for frequencies above 2000 Hz and a slight conductive type hearing loss for lower frequencies in the left ear.
One month later she complained of a rapid further deterioration of hearing in the same ear. At this time an audiometric examination showed a 70 dB perceptive loss in the middle frequencies and no responses for 4000 and 8000 Hz. The ENT specialist considered the possibility of multiple sclerosis; examination by the neurologist, however, did not support this view. Three weeks later the hearing in the other (right) ear also suddenly deteriorated. Following this dramatic event, on the 9th of June, the patient was referred to our clinic for further evaluation.

Examinations

The eardrums were normal and so was tubal functioning. An X-ray examination of the petrous bones showed symmetrical internal auditory canals. General and neurological examination showed no abnormalities and absolutely no signs of multiple sclerosis.
Normal blood and spinal fluid values were obtained. Reiter and VDRL tests were negative.

Audiometry

On the 11th of June 1974, the audiogram showed for the right ear a perceptive loss of 90 dB for all frequencies and no speech discrimination at all. The left ear showed a perceptive loss for frequencies below 3000 Hz and no hearing for 4000 and 8000 Hz. Speech audiometry revealed a 50 dB shift of the curve and a maximum speech discrimination of 80%.
Sisi scores were low in both ears, and tone decay tests as well as Bekésy audiograms gave no indication for a retrocochlear lesion. The stapedius reflex was negative in both ears. Four months later, in Oct. '74, the right-ear audiogram was still unchanged, but now the left ear showed a 90-100 dB perceptive loss in the middle frequencies (Fig., upper left). Speech discrimination was absent in both ears.

Vestibulometry

Romberg and Unterberger tests were normal.
An absence of spontaneous nystagmus and on caloric stimulation an unexcitability of both labyrinths was found.

Electrocochleography

As the nature of the hearing loss was completely unknown, electrocochleography was performed twice on the left ear in order to study the localisation of the lesion. The first ECoG was made when there was a 60 dB loss in speech frequencies in subjective audiometry (June '74), the second (Oct. '74) when audiometry showed an almost total hearing loss.
The first electrocochleogram, which was performed in June '74 on the left ear, showed thresholds varying from approximately 45 dB in speech frequencies to 75 dB at 8000 Hz, while the hearing loss in subjective audiometry was 60 dB in speech frequencies and no responses were

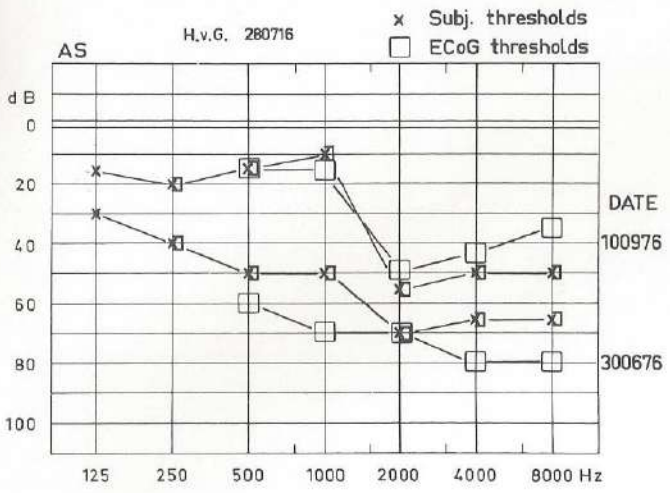
recorded at higher frequencies. So over the whole frequency range ECoG thresholds were 15-20 dB better than subjective thresholds. Input-output curves, with exception of the curve for 500 Hz, all had normal slopes, which means that only for 500 Hz recruitment was likely. Four months later a second electrocochleogram was performed, on the same ear, when subjective audiometry showed an almost total hearing loss (Fig., upper left). For the speech frequencies the ECoG thresholds were 50 dB average, and up to 85 dB at 4000 and 8000 Hz. Now input-output curves showed recruitment for all frequencies (Fig., upper right). Amplitude-latency curves were abnormal at 1500 and 4000 Hz (Fig., lower left). Summating potentials were of normal (4 μV) amplitude and of negative polarity for 1500 Hz, but absent for the higher frequencies. While there was a large difference in subjective thresholds at the time of both electrocochleograms, electrocochleographic thresholds were almost identical to each other.
At the time of the first electrocochleogram the nature of the hearing loss was not understood. Although the difference of 15-20 dB between subjective and ECoG thresholds suggested a retrocochlear lesion, one should realize, that 15-20 dB difference between both thresholds may be within normal limits. A differentiation between neural and cochlear involvement could not be given. The second electrocochleogram, however, which was performed at the time of an almost total hearing loss at subjective audiometry, presented us with much more information. Although thresholds did not differ much from the first electrocochlegram, cochlear damage now became more obvious, which was demonstrated by the presence of recruitment for all frequencies and haircell lesions for 2000 Hz and above. The retrocochlear component also became evident.

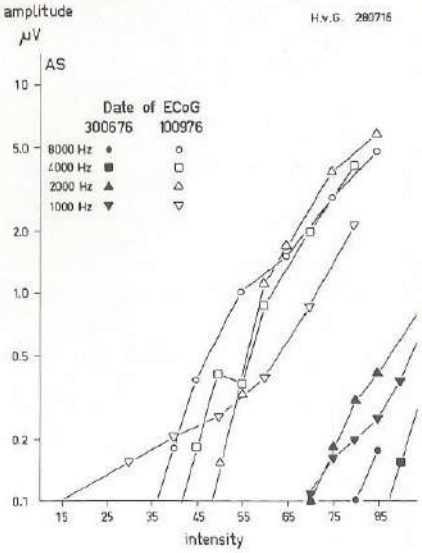
Discussion

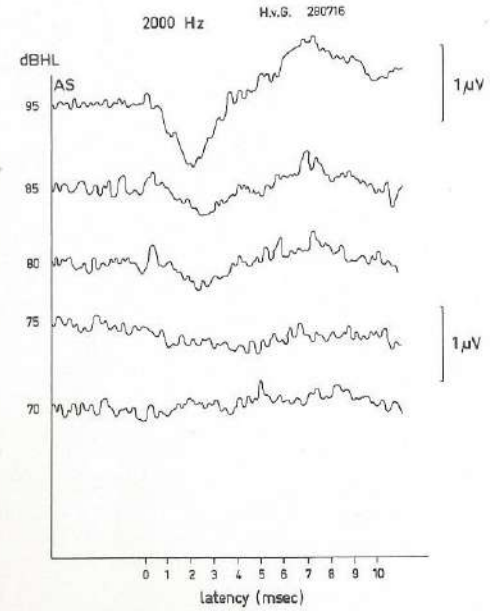
After the first electrocochleogram, which showed no evidence of cochlear damage and suggested the possibility of a retrocochlear lesion, our patient was treated with a vasodilator regimen consisting of large doses of intravenous histamine. No improvement in hearing can be reported as a result of this treatment. The hearing thresholds showed large variations in time, varying from a 40 dB loss to a total hearing loss for both ears.
Thereafter, treatment with steroids was tried, also without succes (it must be noted that these therapies were experimental, since the cause of the disease still remained unknown).
A hearing aid was fitted at the right ear. At present the hearing loss is still fluctuating, while the possible underlying systemic disease is still unknown.

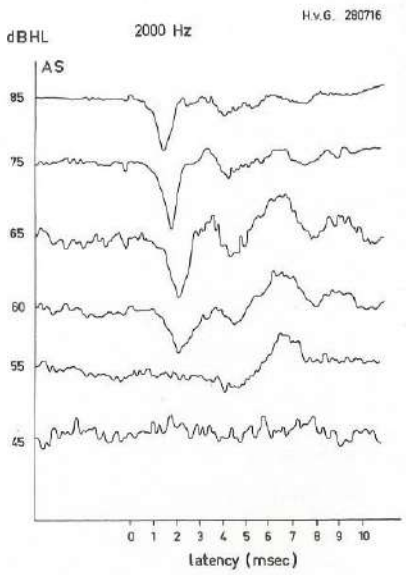
When electrocochleographic thresholds are better than those found in subjective audiometry, this is indicative for a retrocochlear or central component of the hearing loss.
While the first electrocochleogram indicated a sensorineural disturbance without recruitment, the presence of recruitment became evident in the second electrocochleogram. However, thresholds in both electrocochleograms were about the same while subjective thresholds differed largely.

In conclusion, although the final diagnosis remains obscure, the electrocochleographic results indicate a lesion of the cochlea and a fluctuating retrocochlear or central lesion.









B 3.17 Recovery from nearly complete sudden deafness

Case nr. 280716

History

A 60 year old male patient visited our hospital two weeks after he suddenly became deaf in his left ear. Tinnitus was constantly present, but he did not complain of dizziness or vertigo. He had suffered from a myocardial infarction two years ago since when he had been continuously using anticoagulant drugs.

Examinations

No abnormalities at the eardrums were found. Tubal functioning was normal.
General physical and neurological examination showed a healthy man, except for his myocardial status. Blood and spinal fluid chemistry was normal. X-rays of the petrous bones showed normal internal ear canals.

Audiometry

Pure tone audiograms demonstrated normal thresholds at the right ear, but an almost total pure perceptive loss at the left side. Thresholds were approximately 100 dB for speech frequencies.

Vestibulometry

The positional tests were normal.
A minimal positional nystagmus to the right side was found, but no other abnormalities became apparent.

Indications for electrocochleography

Electrocochleography was indicated to investigate a cochlear or retrocochlear cause of the hearing loss. In case of a purely cochlear cause, it is important to know to which extent outer haircell lesions are present. Treatment with a vasodilator intravenously was planned to be started after the electrocochleogram in case a cochlear hearing loss was found.

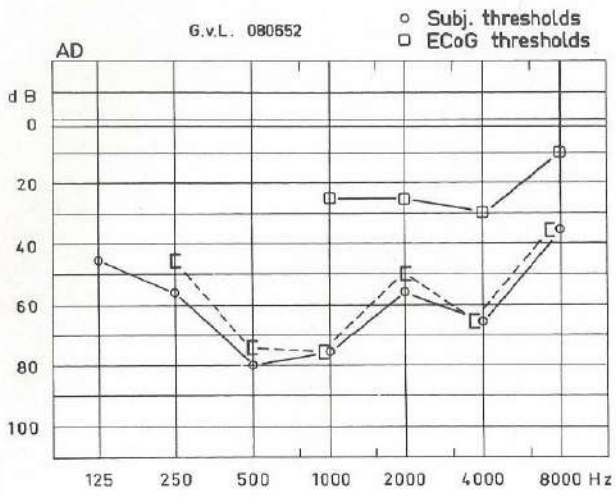
Results of electrocochleography and further convential audiometry

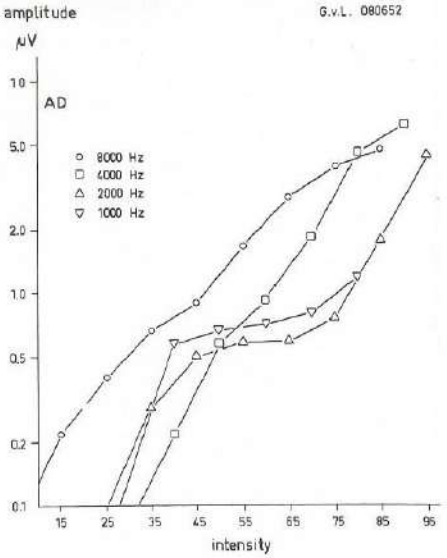
The first electrocochleogram (of which no diagrams are shown) of the left ear showed an average hearing loss of 85 dB. No summating potentials were present for the standard intensities (85 dB – 2000 Hz, 80 dB – 4000 Hz and 85 dB – 8000 Hz), indicating severe haircell malfunctioning. However, at a 100 dB level, at 4000 Hz a small negative summating potential was present, having an amplitude of $-0.4\ \mu V$.
Although on the basis of the electrocochleogram the prognosis of the hearing loss seemed to be discouraging, treatment with intravenously given vasodilators was started. Each day a pure tone audiogram was made; thresholds remained unchanged until the fifth day, on which a 60 dB average hearing loss for speech frequencies was recorded. The speech audiogram showed a 70 dB shift of the curve, with a maximum discrimination score of 50%. Thresholds of the second electrocochleogram (fig. upper left) showed a 70 dB average loss, 10 dB worse than the subjective audiogram. Input-output curves (Fig., upper right, black symbols) de-

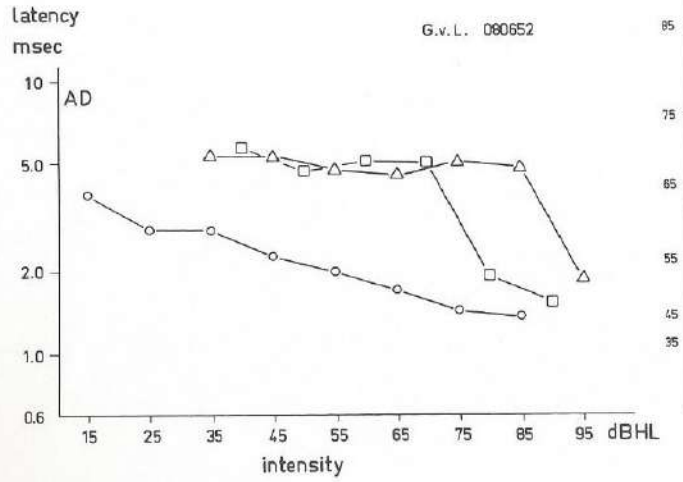
monstrated recruitment for all frequencies. This time also summating potentials were absent for standard intensities. Again a negative summating potential at 4000 Hz and 100 dB intensity was recorded. However, compared with the former electrocochleogram the amplitude was smaller: $-0.2\ \mu V$ instead of $-0.4\ \mu V$. The same conclusion was made: cochlear hearing loss and damage of the outer haircells in the basal turn.

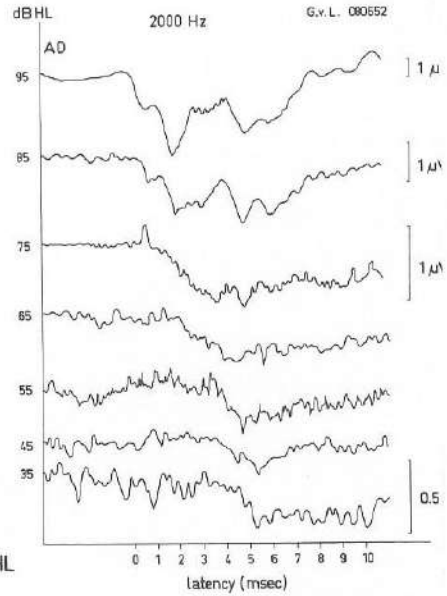
Discussion

The administration of vasodilators was continued for two weeks. The pure tone audiogram, made on the day the patient left the hospital, demonstrated a 45 dB average loss for speech frequencies, indicating further improvement. Two months after being hospitalized, the pure tone audiogram (Fig., upper left, 100976) showed almost normal thresholds up to 1500 Hz and a 50 dB average perceptive loss from 2000 to 8000 Hz. Because of this rather surprising improvement, which was not expected on basis of the previous electrocochleograms, a third electrocochleogram was made. Thresholds corresponded well with those found in subjective audiometry. Recruitment had disappeared for 500 and 1000 Hz, along with threshold improvement. Although, compared with the previous electrocochleogram, thresholds for 2000, 4000 and 8000 Hz also had improved (but less), recruitment for these frequencies remained present. Most important, however, was the presence of summating potentials. The polarity was negative for 2, 4 and 8 kHz amplitudes respectively being $-0.75\ \mu V$, $-0.75\ \mu V$ and $-1.1\ \mu V$ at standard intensities (being low 'normal'). This time, the following conclusion was made:
Cochlear hearing loss, with recruitment for higher frequencies, and diffuse outer haircell lesions in the basal turn.
In this case of sudden deafness the prognosis of the irreversibility of the hearing loss, stated on basis of the first electrocochleogram, has not been proven accurate for the ear as a whole, and only partially for the basal turn.









B 3.18 Retrocochlear hearing loss due to compression of the auditory nerve

Case nr. 080652

History

A previously healthy, 24 year old female patient suddenly became deaf in her right ear. No tinnitus or vertigo was present, middle ear infections had not occurred.
No relatives of the patient were hard of hearing.

Examinations

The aspect of both eardrums was normal. On gross examination no nystagmus was seen, the Romberg and Unterberger tests were normal. The function of the facial nerve was also normal. Röntgenography of the petrous bones showed heavy sclerosis all over both bones. The inner ear canals were extremely narrowed by sclerosis to a width of 2 millimeters.

Audiometry

Pure tone audiometry of the left ear showed normal thresholds, but a 20 dB shift of the speech audiometry curve with respect to normal. A maximum discrimination of 100% was reached. The right ear was much more severely affected. A perceptive hearing loss of 70 dB average for speech frequencies was found (fig. upper left) and discrimination of speech appeared to be totally absent.
The tone decay tests and the Békésy audiogram (Type IV) were abnormal, indicating a retrocochlear origin of the hearing loss. Impedance measurements were normal, but the stapedius-reflex was absent in the right ear.

Vestibulometry

No spontaneous nystagmus was found. Both labyrinths did not respond to caloric stimuli, not even with ice-water. No central symptoms were present. These results are suggestive for a long existing, slowly progressive disorder of both labyrinths.

Indications for electrocochleography

Although the cause of the hearing loss and loss of vestibular function was already revealed by röntgenography, we considered it of importance to know whether the hearing loss was solely the result of nerve compression or that cochlear disturbances were involved too.

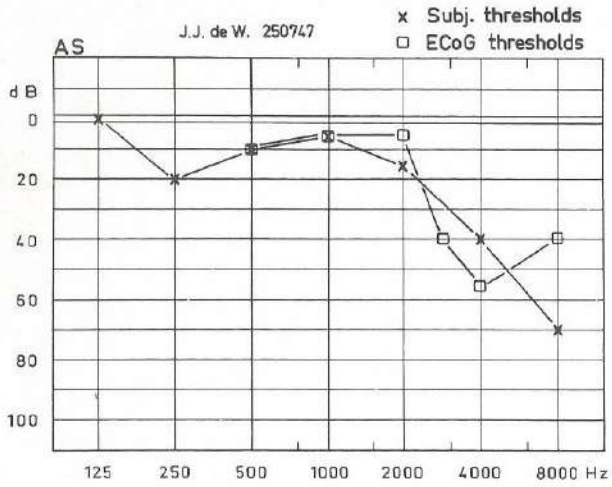
Results of electrocochleography

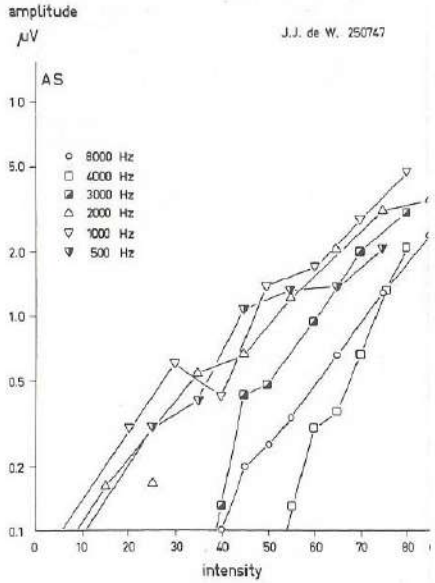
Electrocochleography was performed on the right ear.
Thresholds were far better than those found in subjective audiometry (Fig., upper left). The input-output function was normal for all frequencies recorded, indicating that no recruitment was present. Latency-intensity curves (Fig., lower left) were abnormal for 2000 and 4000 Hz. For these frequencies a sudden latency shift is seen at 95 and 80 dB respectively. The waveform (Fig., lower right) demonstrated a typical feature which is usually found in pontine angle tumors: a prominent deflection with a latency of approximately 5 milliseconds, which persists while intensity decreases.
Narrow-band analysis showed monophasic action potentials.

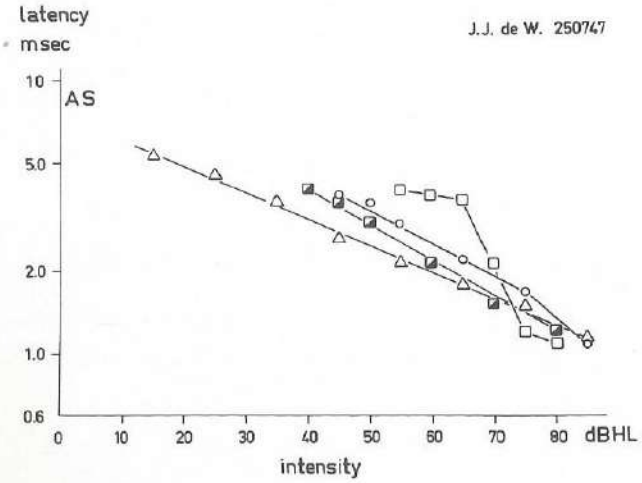
It was concluded that cochlear function was normal (SP-amplitude: $-0.85\ \mu V$ at 85 dB, 2000 Hz) and that the threshold elevation was due to a neural loss.

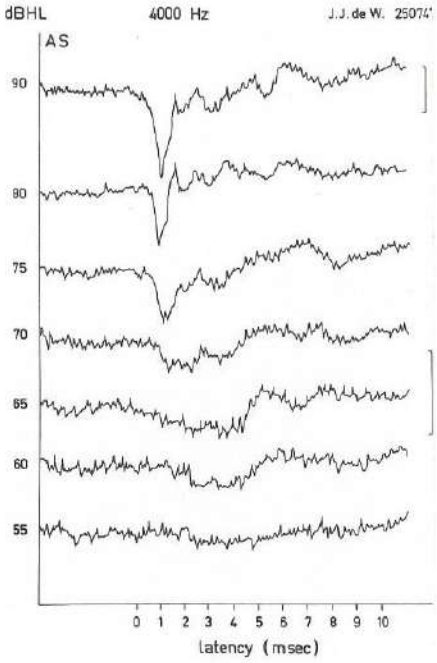
Discussion

Electrocochleography demonstrated a lesion of the auditory nerve.
On the basis of the results of the X-rays one must differentiate between several sclerosing processes of bone: e.g. osteopetrosis and fibrous dysplasia. The cochlear and vestibular nerve are already affected by the disease, but the facial nerve is still normal at both sides. A decompression of the facial nerve seems indicated.









B 4.19 Hearing loss due to gentamycin

Case nr. 250747

History

A 26 year old male patient, complaining of bilateral hearing impairment and tinnitus, was referred to our clinic by his pneumonologist. He had suffered from hearing loss and tinnitus for three months after a six week treatment with gentamycin (3 times 80 mgrs daily) for a serious infection of the lungs caused by pseudomonas aeruginosa. No vertigo had been noticed by the patient.

Examinations

The aspect of the eardrums was normal, as was tubal functioning.

Audiometry

Pure tone audiometry demonstrated a symmetrical perceptive hearing loss. In both ears thresholds were almost normal up to 2000 Hz, above 2000 Hz there was a sharp decrease to a loss of ± 50 dB for 4000 and 8000 Hz (Fig., upper left). Speech audiometry showed a 15 dB shift of the curve, but a maximum speech discrimination of 100% was reached at both ears at sufficient amplification. Sisi-scores were high (90-100%) for 3000, 4000 and 8000 Hz. The tone decay test was normal.

Vestibulometry

The positional tests (Romberg, Unterberger) were normal.
A minor nystagmus to the left side was seen in all head and body positions.

Indications for electrocochleography

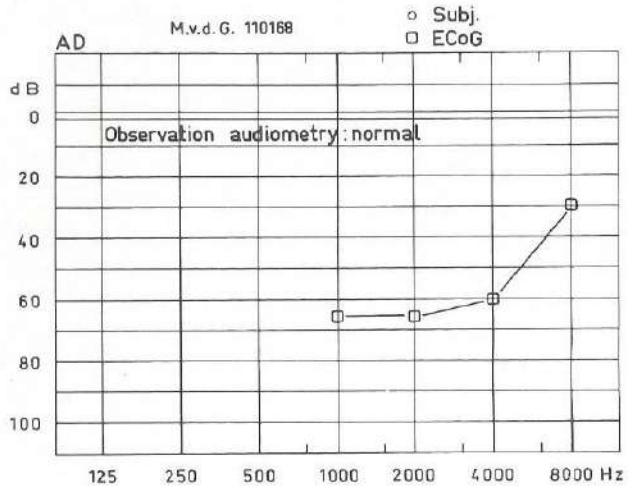
On the basis of the results from subjective audiometry, a cochlear hearing loss was very likely. In the case of a pure cochlear hearing loss it would be interesting to know if irriversible haircell damage was present.

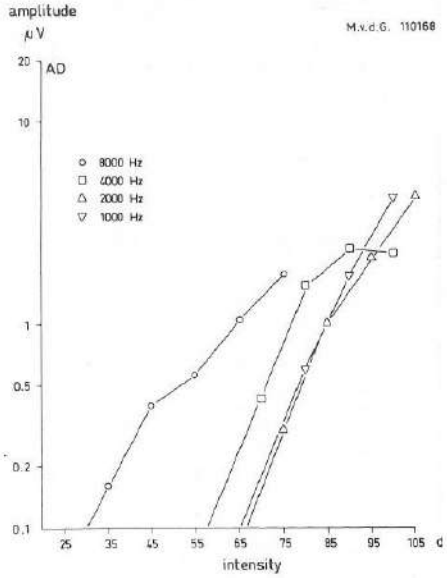
Results of electrocochleography

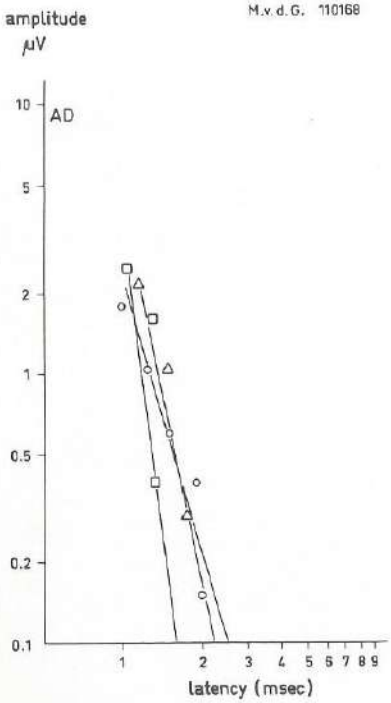
Electrocochleography was performed on the left ear.
Thresholds confirmed those found in subjective audiometry (Fig., upper left). Input-output curves showed no clear recruitment for 3000 and 4000 Hz, but the curves were steeper than the curve for 2000 Hz, for which recruitment clearly was absent. The amplitude-latency curve was also normal for 2000 Hz, but abnormal for the higher frequencies. Summating potentials were of negative polarity for all recorded frequencies, the amplitudes being abnormally small for 4000 and 8000 Hz ($-0.40\ \mu V$, 80 dB and $-0.30\ \mu V$, 85 dB respectively), and normal for 2000 and 3000 Hz ($-1.35\ \mu V$, 85 dB and $0.70\ \mu V$ 80 dB respectively). It was concluded that the hearing loss was of cochlear origin with recruitment for 3000 and 4000 Hz (possibly also for 8000 Hz) and outer haircells lesions for 3000, 4000 and 8000 Hz.

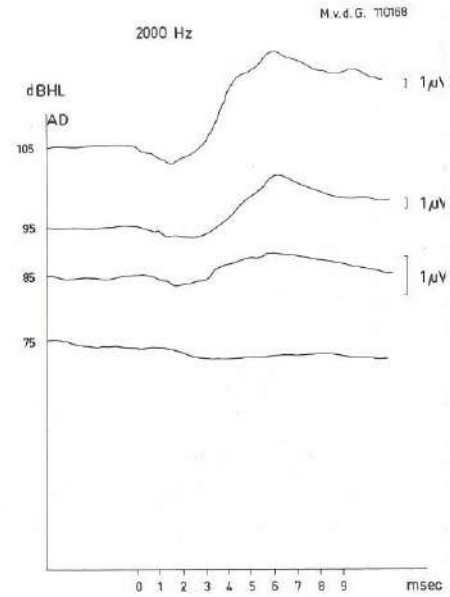
Discussion

The high-frequency perceptive hearing loss was already known from subjective audiometry. ECoG demonstrated the functional loss of outer-haircells in the basal turn. The tinnitus was treated with vasodilator drugs, but this treatment has not been succesful.
The pure tone audiogram has not improved, as was expected from the electrocochleographic results.









B 4.20 Hearing evaluation of a child with retardation of speech and language

Case nr. 110168

History

A five year old boy, born seven weeks prematurely, was sent to our clinic because of considerable retardation of speech and language. At the time of his birth the child had been blue and artificial ventilation had been given. His motor development seemed normal; however, the first signs of speech did not appear until the age of four. The parents began to doubt the child's ability to hear because on occasion they seemed unable to communicate with the boy.

Examinations

Eardrums and tubal functioning were normal. Neurological examination showed normal motor development and social behaviour. The intelligence quotient was average. At phoniatric examination the child's small vocabulary was striking, and he was not able to produce a sentence of more than two words. Articulation also proved to be bad.

Audiometry

It was not possible to obtain a normal pure-tone audiogram. Therefore, three different examiners performed observation audiometry. Two of them concluded that the child's hearing was normal, while the third examiner had some doubts.

Indications for electrocochleography

Since a hearing loss as the cause for speech retardation could not be excluded with certainty by observation audiometry, it was decided to perform electrocochleography to definitely establish the child's hearing ability.

Results of electrocochleography

Surprisingly, electrocochleography showed a bilateral perceptive hearing loss of about 65 dB in speech frequencies and of about 30 dB at 8000 Hz. The right ear seemed to be the better one.
On basis of the slope of the input-output curves (Fig., upper right) recruitment was not likely to be present for 8000 Hz, while indications for recruitment existed for 1000, 2000 and 4000 Hz. Amplitude-latency curves were abnormal for 2000, 4000 and 8000 Hz. Summating potentials had very small amplitudes.
It was concluded that the hearing loss was of cochlear origin with outer haircell loss except for 8000 Hz, and possible also some neural involvement.

Discussion

High frequencies are always present in the sound producing toys used in observation audiometry. Whereas in electrocochleography high frequency thresholds were better than low and middle frequency thresholds, the favorable results of the observation audiometry could be explained.
As a result of electrocochleography, the cause of the speech and language retardation was now known to be a serious bilateral perceptive hearing loss. Rehabilitation of speech and hea-

ring could be started. A hearing-aid was applied and speech-therapy was intensively given,which resulted in a rapid improvement of speech.
When the boy reached the age when normal pure tone audiograms could be made, electrocochleographic thresholds were confirmed for both ears. For the left ear the subjective audiogram was better at the lower frequencies by 15 dB, but it also corresponded well at the higher frequencies.

VI Discussion

Performing electrocochleography in children generally answers the question about the validity of their auditory periphery (Spoor and Eggermont, 1976). In adults, it gives cues in suspected malingerers or patients simulating some kind of deafness. In these case findings of normal peripheral hearing do not exclude the presence of some hearing problem. They exclude, however, the presence of a conductive or cochlear hearing loss, e.g. noise trauma. The reliability of ECoG is partly due to the fact that it is not influenced by sedation or general anaesthesia and cannot be influenced by attention either. This is in contrast with the slow vertex potential (in use in the late sixtees and early seventies) which is influenced by attention, sleep and sedation and which is absent under general anaesthesia (Picton et al., 1974a, b).

VI I The selected examples

a) *Children in which observation audiometry is impossible or unreliable.* (A.1.1. through A.1.5)

This selection gives an impression of the types of children which are 'difficult-to-test'. Examples are given on: the autistic child, the child with conductive losses in which a perceptive loss could not be excluded on basis of behavioural tests, and the child in which subjective audiometry could not differentiate between the relative components in a mixed hearing loss. Spoor and Eggermont (1976) demonstrated that, in 95% of the cases the difference between ECoG thresholds and subjective thresholds can be expected to be less than 20 dB. A hearing loss of less than 20 dB, whether it is conductive or perceptive, therefore cannot be detected by ECoG. In conductive hearing loss a shift to longer latencies is usually seen (Berlin et al., 1973), while the amplitudes of the SP's tend to be smaller. However, of both parameters the spread of normal values covers a range of approximately 20 dB (Eggermont, 1976). This explains the difficulties in determining the relative components in a mixed hearing loss of less than 40 dB (A.1.4, A.1.5). Apart from tympanometry and stapedius reflex tests, bone-conduction electrocochleography would be extremely helpful in these cases. However, calibration problems and artefacts due to mechanical vibrations of the skull have so far prohibited the routine use of a bone conductor in ECoG (Arlinger and Kylén, 1977).

b) *Adults suspected from aggravation or simulation* (A 2.6 through A 2.8)

In these cases, findings of normal peripheral hearing do not exclude the presence of some hearing problem.

However, the use of ECoG offers the way to proceed with the behavioural tests, (e.g. Lombard or Stenger test) although the stapedius reflex-test may provide some information. For instance, in A.2.8 a striking difference between ECoG and subjective thresholds was found.

A far larger group of patients comprises those of which besides the audiogram considerable information is available about the type of hearing loss, e.g. conductive versus perceptive, cochlear versus retrocochlear. However, seldom do all subjective tests agree, and the need for an objective test is often felt.

c) *The validity of hair-cells in cochlear hearing loss* (B 1.9 through B 1.11)

It has been demonstrated in the section on the summating potential that the SP can be regarded as the only reliable parameter at this moment, for estimating hair-cell validity in the basal turn of the cochlea. In Menière's disease a tendency exists to larger SP amplitudes at 2000 Hz compared with normal ears (Aran and Negrevergne, 1973; Schmidt et al., 1974). Small SP amplitudes, a positive SP and especially the absence of SP, indicate severe outer hair-cell dysfunction or anatomical hair-cell loss. In Menière's disease the loss of SP may indicate the change from a fluctuating to a more stable state of the disorder. This knowledge might influence the clinician's choice of therapy.

d) *Diagnosing retrocochlear types of hearing loss* (B 2.12 through B 2.14)

Electrocochleographic thresholds in retrocochlear hearing loss are in general better than or equal to subjective thresholds, since ECoG describes only the auditory periphery (cochlea and peripheral part of the auditory neuron): Innitzer, 1976, Morrison et al. 1976). The degree of threshold difference depends on the site, size and consistency of the process (e.g. beginning or advanced pontine angle tumor), Morrison et al., 1976). A typical waveform of the AP (B.2.14 and B.3.18) is produced when the auditory nerve is compressed or stretched (Beagley et al., 1977, Glazenburg, 1978). This waveform can be examined using the technique of narrow-band analysis (Eggermont, 1976).
The AP waveform found in retrocochlear hearing loss may look similar to the AP found in Menière's disease, which shows a prominent SP^-. Since the SP shows no adaptation, differentiation can often be made on basis of this property.
When retrocochlear hearing loss is due to diffuse lesions in the brainstem, normal input-output curves are found, and, apart from thresholds-differences (B.2.14), the AP-waveform is normal. Besides threshold-differences and broadening of the AP-waveform in retrocochlear hearing loss, Gibson et al. (1976) assign some value to the persistance of the CM. However, no definite conclusion can be derived from the CM, as already has been discussed in the introduction. For more exact information regarding the location of the cause of the retrocochlear hearing loss, brainstem audiometry seems to be extremely useful (Selters and Brackmann, 1977).

e) *Predicting the prognosis in sudden deafness* (B 3.15 through B 3.17)

The most popular concept used to explain 'idiophatic' sudden deafness is viral labyrinthitis and/or vascular occlusion. Lindsay et al. (1960) described degenerative changes consisting of atrophy of the stria vascularis, the tectorial membrane and the organ of Corti in a case of mumps. In such cases it is possible with ECoG to obtain information about the status of the outer hair-cells shortly after the attack. The validity of the hair-cells is represented by the amplitude of the SP, while a change of polarity (from negative to positive) is associated with hair-cell damage. From an electrocochleographic study of 34 patients with sudden deafness Nishida et al. (1976) derived a prognostic criterium for the disease, i.e. dominant negative SPs offer a good prognosis. However, absence of the SP does not always mean a total or permanent loss of outer hair-cell function, as is demonstrated in B.3.17. In neurological diseases the cause of sudden deafness can be retrocochlear. B.3.16 demonstrates a partial retrocochlear origin of sudden deafness, while no other neurological symptoms exist.

VI 2 The clinical significance of electrocochleography

VI 2.1 Diagnosis categories and clinical significance

The clinical significance of ECoG is defined as the influence on the management of hearing disorders.
For the selected examples in general the result of the electrocochleogram in one or more details influenced the optional therapy. The next step has been to investigate this for all the patients on which ECoG was performed from 13-03-71 to 09-11-77. The question to be answered by ECoG on these 531 patients of all ages can be divided in four categories:
A. Is a hearing loss present, and if so, how large is it?
B. If a hearing loss is present, is it a conductive or a perceptive hearing loss?
C. If it is a perceptive hearing loss, is it cochlear or retrocochlear?
D. If it is a cochlear hearing loss, is it a metabolic hearing loss or hair-cell loss?
These four categories are the same as used by Eggermont en Odenthal (1977).

The clinical significance of these 531 electrocochleograms can also be divided into four groups, to be defined as follows:

I Very little prior information was present, the ECoG resulted in a definite diagnosis and thereby strongly influenced therapy.
II Some prior information was present, the ECoG provided significant information, thereby contributing to the diagnosis, and influencing therapy.
III Some prior information was present, the ECoG provided additional information, thereby refining the diagnosis, but not influencing therapy.
IV Much prior information was present, the ECoG results provided no additional information, thereby not influencing therapy.

Table I: The relation between the indication for ECoG and the clinical significance for patients of all ages.

All patients

INDICATION FOR ECoG \ CLINICAL SIGNIFICANCE OF ECoG	I	II	III	IV	total
A (Audiometry)	148	24	21	1	194
B (Conduction loss or perceptive hearing loss)	5	4	1	2	12
C (Peripheral or central hearing loss)	12	62	30	14	118
D (Metabolic dysfunction or haircell loss)	11	22	150	24	207
	176	112	202	41	**531**

Table II: The relation between the indication for ECoG and the clinical significance of ECoG for patients under 13 years.

patients < 13 years

INDICATION FOR ECoG \ CLINICAL SIGNIFICANCE OF ECoG	I	II	III	IV	
A (Audiometry)	128	17	11	0	156
B (Conduction loss perceptive hearing loss)	0	2	1	0	3
C (Peripheral or central hearing loss)	0	0	0	0	0
D (Metabolic dysfunction or haircell loss)	o	1	1	0	2
	128	20	13	0	**161**

Table III: The relation between the indication for ECoG and the clinical significance of the ECoG results for patients over 13 years.

patients ≥ 13 years

INDICATION FOR ECoG \ CLINICAL SIGNIFICANCE OF ECoG	I	II	III	IV	
A (Audiometry)	20	7	10	1	38
B (Conduction loss perceptive hearing loss)	5	2	0	2	9
C (Peripheral or central hearing loss)	12	62	30	14	118
D (Metabolic dysfunction or haircell loss)	11	21	149	24	205
	48	92	189	41	**370**

The results are given in Table I.

If one does not know much more than 'the child is hard of hearing' or the child only shows autistic behaviour, ECoG nearly always has a major impact on the treatment. 148 cases out of 194 in which the basis concern was the need for a detailed audiogram, fall into group I while in just one case it did not help either diagnosis or treatment.
On the other hand, if the patient's audiogram and the presence of recruitment etc. is known, and the question is to distinguish between a metabolic disturbance (cochlear loss with recruitment, no haircell dysfunction) or haircell loss (also with recruitment) 150 out of 207 cases resulted in a refinement of diagnosis, but with no influence on the therapy.

Using correlation techniques for grouped data (Spiegel, 1961), the correlation coefficient between the rankings of clinical significance (I to IV) and the degree of refinement added by performing ECoG (A to D) has been calculated to be $r = 0.67$. This is significantly different from zero ($p < 0.0005$); the 95% confidence limits of r are between 0.62 and 0.71. It indicates a moderately high positive correlation between the degree of refinement in the diagnosis asked for, and the decrease in the clinical significance of the ECoG result.
When the total group of patients is divided in a group of children up to and including 13 years, and a group of patients above this age, the results are different as is il-

lustrated in Table II and Table III. The criteria are the same as for the group as a whole. It must be noted that usually children require general anaesthesia, while in adults only a premedication is necessary. Therefore, in deciding on performing ECoG in children, the chance of obtaining rewarding information should be high. For patients older than 13 years mostly a differentiation between metabolic disturbance and hair-cell loss was sought. The relatively large number of patients in this group is the result of the special interest of the Leyden E.N.T. clinic in Menière's disease. In 48 out of 270 patients the diagnosis could not have been made without ECoG results (18%), while in 34% (92 out of 270) ECoG has been helpful in obtaining diagnosis. These results contrast with the results from the children tested: in 128 out of 161 patients diagnosis has been made by ECoG only (80%), while in 12% (17 out 161) ECoG has been helpful in establishing diagnosis.
It seems that electrocochleography is clinically significant in obtaining exact information about hearing thresholds in children. This is not necessarily true in adults since a reliable subjective audiogram can generally be obtained. Thus, the more refined the question to be answered by ECoG, the less significant are the results for the clinician.

VI 2.2 Comparison of the Leyden results to a worldwide questionnaire

Elaborating on a questionnaire distributed throughout the world, Crowley et al. (1975) asked for a similar comparison for 3180 patients. Part of this group could be separated in 553 adults and 871 children under 15 years. Their definitions of significance are in global accordance with the groups I through IV in this investigation:

I Diagnosis very doubtful, ECoG primary factor in diagnosis
II Diagnosis in some doubt, ECoG added useful information
III Diagnosis clear pre-ECoG, confirmation desired
IV Diagnosis remained doubtful, in spite of ECoG

A comparison shows that the Leyden results and the results from the Crowley questionnaire for all patients are significantly different (X^2.995). Differences may be the result of the slightly different definitions, but also of a different selection of the patients submitted to ECoG.
Comparison of the adult groups results in an apparently better result (12.3%) for the Leyden group in category I, which also may be caused by the above mentioned variables and the fact that except for the Leyden ECoG's, the ECoG's in the Crowley questionnaire were made with clicks (no reliable audiograms can be obtained with clicks) (Table V).
The children-group is shown in Table VI. Here the percentage in category I is again significantly higher for the Leyden group, but adding the percentages of category I and II gives percentages of 92 for Leyden and 87.8 for the above mentioned questionnaire respectively.
It is not surprising that the patient material in the present study and in the Crowley questionnaire is different. Patients to be subjected to ECoG are selected on diverging grounds.
One group, mainly children, is selected because no other tests provide enough information and it is therefore likely that the ECoG results will be classified in groups I

or II. Another group, e.g. patients suffering from Menière's disease, is selected for the main purpose of studying this less understood disease. It is therefore not suprising that the results obtained would add anything to the choice of the mostly trial-and-error-type therapy. Most of these patient results are therefore classified in Groups III or IV. Patients not belonging to the children or Menière group are not frequently found because either the direct consequences are minimal and no serious complaints about the hearing loss are present, with e.g. the exception of a few cases of sudden deafness. But they belong to the same general category as Menière's disease: a poorly understood problem. Nevertheless, our results are indicative for the application of ECoG and it may be concluded, that for about 90% of the children who were examined with ECoG, the ECoG result added useful to significant information to a doubtful diagnosis. In our population of adults the clinical significance of ECoG (category I and II) is still present in about 30% of the cases, while in 50-60% diagnosis can be confirmed and refined, which is surely a positive contribution to the investigation of hearing disorders.

VI 2.3 Conclusion

The investigation in this thesis has dealt mainly with the practical clinical significance of electrocochleography, but it may well be that at the present time the diagnostic possibilities extend beyond those of therapy. One of the most promising features consists in studying various cochlear disorders, such as e.g. Menière's disease or sudden deafness. The method provides information about diseases that, at present, cannot be obtained by other methods. This ultimately may result in changes in the management of these disorders, and even in causal therapy.

Table IV: Comparison of the Crowley questionnaire and the Leyden results

CLINICAL SIGNIFICANCE OF ECoG / ALL PATIENTS	I	II	III	IV	
LEYDEN (531)	33	21	38	8	%
CROWLEY et al. (3180)	22.3	31.6	43.9	2.2	%

Table V: Comparison of the Crowley questionnaire and the Leyden results for adults

ADULTS / CLINICAL SIGNIFICANCE OF ECoG	I	II	III	IV	
LEYDEN 370	12.3	25	51	11	%
CROWLEY et al. 553	2.0	32.2	65.3	0.5	%

Table VI: Comparison of the Crowley questionnaire and the Leyden results for-children

CHILDREN / CLINICAL SIGNIFICANCE OF ECoG	I	II	III	IV	
LEYDEN (161)	79.5	12.5	8	0	%
CROWLEY et al. (871)	48.2	39.6	11.3	0.9	%

VII Summary

Electrocochleography (ECoG) is the recording of sound-evoked electrical potentials from the human inner ear and the auditory nerve. This method can be used to determine the hearing ability of the patient. To compare the results of this objective method with the results of subjective audiometry, frequency specific sound stimuli (short-tone-bursts) are used in electrocochleography. In the absence of a central component of the hearing loss, electrocochleography can give definite information regarding a) the localization of the (peripheral) hearing loss, b) the nature of the hearing loss, and c) a more precise differentiation of the cochlear damage.
The goal of this investigation is to illustrate and evaluate the clinical significance of the method, using case studies of 20 selected patients, and the influence of the electrocochleographic results on the management of the hearing disorders, as investigated on basis of all electrocochleograms performed in Leyden (531 in a six year period) (Chapter I).

The recorded response to tonebursts presented alternately in phase consists of a mixture of summating potential (SP) and action potential (AP), both of which are quantified by their amplitude values. In addition, the latency of the AP is taken into account. The AP threshold intensity corresponds to the subjective hearing threshold in cases of peripheral hearing loss. The supraliminal behaviour of the auditory periphery is quantified by various stimulus-response relationships (e.g. AP-amplitude-intensity, latency-intensity), as well as by the amplitude of the SP at a fixed stimulus intensity. This quantification permits a comparison with some subjective measures, e.g. the amount of recruitment in relation to the steepness of the AP-amplitude-intensity curve (Chapter II).

The promontory recorded summating potential is mainly generated in the outer hair cells of the basal turn of the cochlea. In case of non-functioning or absence of part of the hair cells, the amplitude of the summating potential decreases, while in some cases, at high-frequency stimulation, the polarity changes from negative to positive. The envelope of the sound-induced excitation pattern along the cochlear partition consists of a rising part located more near the basal turn of the cochlea (producing an SP^-), and a falling part located more apically (producing an SP^+).
In cochleas with hair-cell loss the contributions of the various regions can be changed in such a way, that less SP is produced, resulting in a decrease of SP^- and therefore in a change of polarity of the recorded SP from SP^- to SP^+. (Chapter III).

Different types of hearing loss generally produce different electrocochleographic characteristics.

a. A conductive hearing loss causes threshold elevation and a tendency to larger latencies of the AP and smaller AP and SP amplitudes.
b. A cochlear hearing loss causes threshold elevation, but the AP amplitude at high intensities remains the same as in normal ears (recruitment). Absent or positive summating potentials indicate outer-hair cell loss (functional or anatomical).
c. In central (retrocochlear) hearing loss electrocochleographic thresholds are often better than subjective thresholds. The AP-waveform may be changed, which results in longer latencies at intermediate intensities as compared to normal ears (Chapter IV).

Twenty selected patients are extensively described; a division is made in five groups with regard to their clinical problem:
A1 Observation audiometry is impossible or unreliable.
A2 Suspicion for aggravation or simulation.
B1 Estimation of hair cell validity in cochlear hearing loss (e.g. Menière's disease).
B2 Diagnosing retrocochlear types of hearing loss (e.g. pontine angle neuroma).
B3 Predicting the prognosis in sudden deafness. (Chapter V).

A survey of 531 electrocochleograms performed in Leyden over a period of 6 years is used for evaluating the clinical significance of ECoG. The clinical significance of the ECoG test ranges from 'no refinement of the diagnosis could be made' to 'without ECoG no diagnosis could have been made and any therapy given would have been without basis'. A ranking of this implication into four categories was made. The ultimate indication for performing the test is also ranked into four categories from 'nothing is known about the hearing ability' to 'a refinement about the type of cochlear hearing loss is asked for'.
For the group as a whole a moderately high negative correlation was found between the degree of refinement asked for and the clinical significance of the test.
In children up to and including 13 years it was seen that ECoG added significant information to the clinical picture in 90% of the cases, while in patients older than 13 years this is reduced to 30%. (Chapter VI).

Samenvatting

Electrocochleografie (ECoG) is de registratie van door geluid opgewekte elektrische potentialen in het binnenoor en de gehoorzenuw. Deze methode kan worden gebruikt voor de bepaling van gehoorsfunktie. Om de resultaten van deze objektieve methode te kunnen vergelijken met subjektieve vormen van gehooronderzoek, wordt ook bij electrocochleografie gebruik gemaakt van frekwentie specifieke geluidsstimuli: korte toonstoten.
Behalve bij centrale gehoorverliezen kan electrocochleografie belangrijke informatie geven over: a) de plaats van het (perifere) gehoorverlies, b) de aard van het gehoorverlies, en c) nadere differentiëring van cochleaire beschadigingen.
Het doel van dit onderzoek is het evalueren van de klinische betekenis van electrocochleografie, waarbij ter illustratie gebruik gemaakt wordt van de ziektebeschrijving van 20 geselecteerde patienten. De invloed van de resultaten van 531 in Leiden verrichte electrocochleografieën (gedurende een periode van 6 jaar) op het medisch handelen wordt onderzocht. (Hoofdstuk I).

De geregistreerde respons op korte toonstoten, welke met alternerende fase worden aangeboden, is samengesteld uit de sommatiepotentiaal (SP) en de actiepotentiaal (AP); beide worden gekwantificeerd door de waarden van hun amplitude. De latentie van de AP is eveneens van belang. De intensiteit van de AP drempel komt bij perifere gehoorverliezen goed overeen met de subjektieve hoordrempel. De bovendrempelige verschijnselen in het perifere auditieve systeem kunnen worden gekwantificeerd door verschillende stimulus-response relaties (b.v. AP-amplitude-intensiteit, latentie-intensiteit) en door de amplitude van de SP bij een vaste intensiteit. Door deze kwantificering wordt een vergelijking met bepaalde subjektieve symptomen mogelijk, zoals b.v. de mate van recruitment en de steilheid van de AP-amplitude-intensiteitscurve. (Hoofdstuk II).

De van het promontorium afgeleide sommatiepotentiaal wordt hoofdzakelijk gegenereerd in de buitenste haarcellen van de basale winding van de cochlea. Wanneer een deel van de haarcellen niet funktioneert, neemt de amplitude van de SP af; in sommige gevallen, bij hoogfrequente stimulatie, kan de polariteit van de SP omkeren van negatief naar positief. De omhullende van het (door geluid opgewekte) excitatieprofiel in de cochlea bestaat uit een stijgend deel, dat ter plaatse van de basale winding van de cochlea is gelegen, en een negatieve SP produceert, en uit een dalend deel, meer naar de apex van de cochlea gelegen, dat een positieve SP produceert.
Wanneer er haarcel verlies in de cochlea bestaat, kunnen de bijdragen van de verschillende gedeelten van de cochlea zo veranderen, dat er minder SP^- wordt geproduceerd, hetgeen resulteert in een verkleining van de amplitude van de SP^-. Dit kan

weer leiden tot een verandering van de polariteit van SP^- naar SP^+. (Hoofdstuk III).

Verschillende typen gehoorverlies hebben in het algemeen ook electrocochleografisch verschillende eigenschappen.
a) een *geleidingsverlies* uit zich in drempelverhoging, en een neiging tot langere latenties voor de AP en kleinere amplituden van de SP en de AP.
b) een *cochleair gehoorverlies* uit zich in drempelverhoging, maar de AP-amplitude blijft bij hoge intensiteiten dezelfde als bij normaal horenden (equivalent met recruitment
c) Bij een *centraal* (retrocochleair) gehoorverlies zijn de met electrocochleografie gemeten drempelwaarden vaak beter dan die gevonden met subjektieve audiometrie. Tevens kan de golfvorm van de AP veranderd zijn, hetgeen vergeleken met normaal horenden tot uitdrukking komt in langere latenties bij middelhoge intensiteiten (Hoofdstuk IV).

Twintig geselecteerde patiënten worden uitvoerig beschreven; deze worden naar de klinische probleemstelling in vijf groepen verdeeld:
A1) Spelaudiometrie is niet mogelijk of onbetrouwbaar.
A2) Verdenking op aggravatie of simulatie.
B1) Het bepalen van de validiteit van de haarcellen bij cochleaire gehoorverliezen (b.v. de ziekte van Menière).
B2) Het diagnostiseren van gehoorverliezen van het retrocochleaire type (b.v. brughoektumoren).
B3) Het geven van een prognose bij plotselinge perceptieve gehoorverliezen. (Hoofdstuk V).

Ter evaluatie van de klinische betekenis van electrocochleografie wordt gebruik gemaakt van een overzicht van 531 electrocochleogrammen welke in een periode van 6 jaar in Leiden zijn verricht. De klinische betekenis van het electrocochleogram varieert van 'de diagnose kon niet verder verfijnd worden' tot 'zonder het electrocochleogram kon geen diagnose worden gesteld en een gegeven behandeling zou ongefundeerd zijn.' Deze klinische betekenis wordt in vier categorieën onderverdeeld, evenals de uiteindelijke indicatie waarvoor het electrocochleogram werd verricht. De indicatie varieert van 'enige kennis omtrent de gehoorsfunktie ontbreekt' tot 'een verfijning van het type cochleair gehoorverlies wordt gevraagd'.
Voor de groep als geheel wordt een matig hoge negatieve correlatie gevonden tussen de mate van verfijning waarom werd gevraagd en de klinische betekenis van het electrocochleogram. Wanneer ECoG werd verricht bij kinderen tot en met 13 jaar, bleek het resultaat van het onderzoek in 90% van de gevallen belangrijke informatie toe te voegen aan het klinische probleem; voor patiënten boven 13 jaar blijft dit aantal beperkt tot 30% (Hoofdstuk VI).

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