

NEURO-OTOLOGICAL ASPECTS OF CEREBELLOPONTINE ANGLE TUMORS

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.

Anatomy

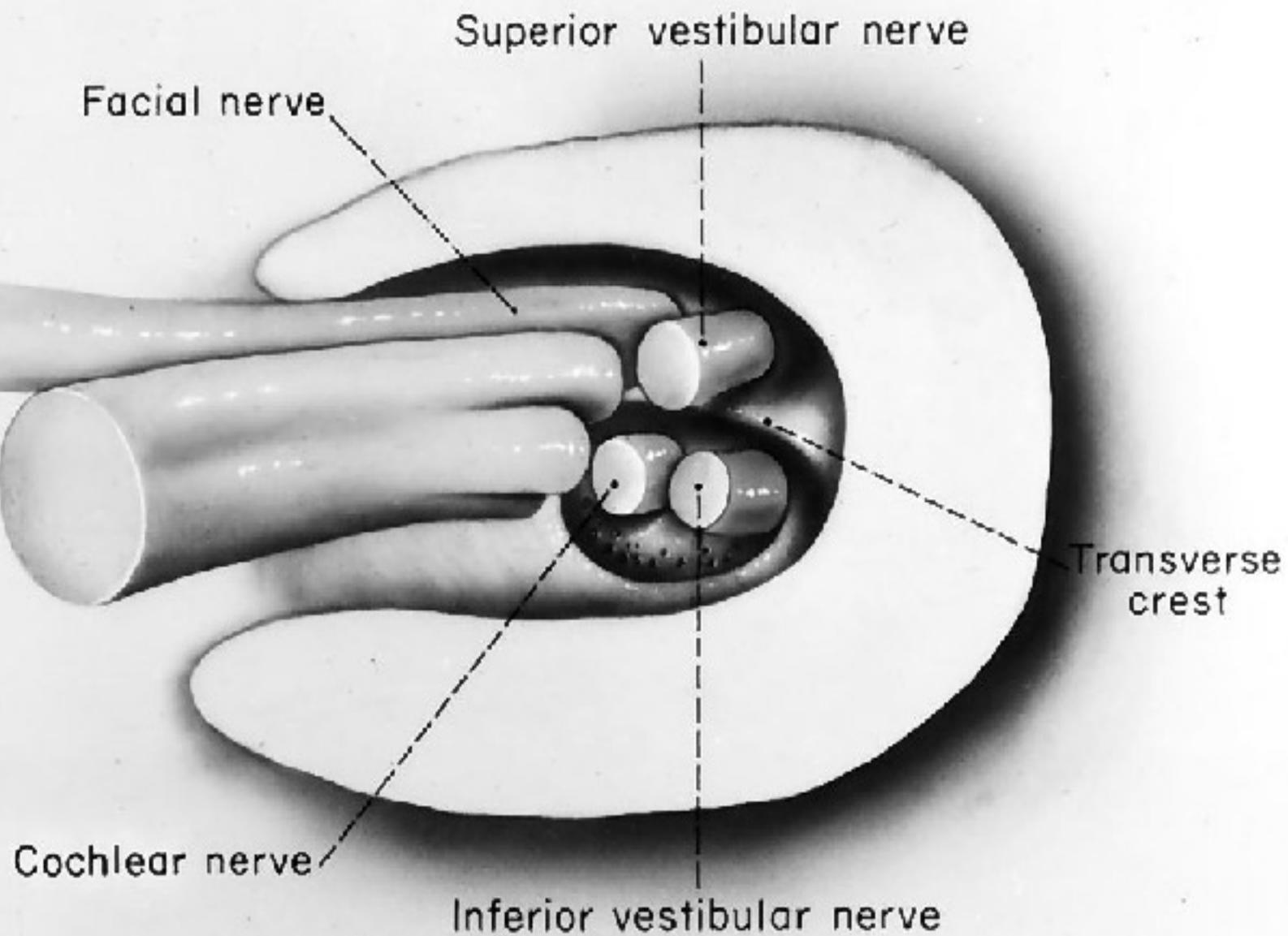
- Boundaries of CP angle
 - Medial
 - Lateral
 - Superior
 - Inferior

Anatomy

Internal acoustic meatus.

- The lateral wall is divided into superior and inferior halves by the falciform crest.
- The upper compartment is separated into an anterior area for the facial nerve and a posterior area for the superior vestibular nerve, by a sharp vertical ridge of bone known as 'Bill's bar' after William House.
- The lower half comprises two areas, anteriorly the tractus spiralis foraminosus through which the cochlear nerve passes, and posteriorly the inferior vestibular nerve supplying the saccule.

A



Anatomy

- Inner ear (labyrinth)
 - Bony labyrinth : Perilymph
 - Membranous labyrinth : Endolymph
- Cochlea
 - 2 $\frac{3}{4}$ turns
 - Scalae
- Organ of Corti
- Semi-circular canals

Neurophysiology

- Auditory pathway
- Hair cells
- Action potentials in auditory nerve fibres

Effects On The Inner Ear

- Cochlear changes may result from interference with the arterial blood supply of the inner ear from pressure of tumour on branches of the internal auditory artery .
- Degeneration is more commonly seen in the cochlea than in the otolith organs or in the semicircular canals.
- Atrophy of the organ of Corti, most frequently seen in the basal turn, but occasionally widespread or complete).
- Vacuolization of the stria vascularis

Clinical Presentation

- Greatest incidence in the fourth, fifth and sixth decades.
- More incidence in females.

Otological Stage Deafness And Tinnitus

- The commonest symptoms are unilateral hearing loss and tinnitus, which occur in over 90% of patients. The deafness is usually gradual in onset and slowly progressive over a period varying from as little as a few months to 20 years or more, but averaging about 2 years
- The patient may volunteer the information that his ability to discriminate speech seems disproportionately poor, especially when conversing on the telephone.
- In perhaps 10% of cases the hearing loss is sudden and may be profound, due presumably to a vascular accident to the cochlea.

Deafness And Tinnitus

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- Fluctuating low frequency hearing loss
- Variations in speech discrimination
- The tinnitus is non-pulsatile, high pitched and ipsilateral to the side of lesion and usually commences at about the same time as, or precedes, the deafness.
- Mechanism of tinnitus in patients with acoustic neuroma is thought to be the same as that for hearing loss

Otological Stage Imbalance

- The slowly growing tumour destroys the vestibular nerve from which it arises so gradually that the central nervous system is able to compensate for the unilateral loss of peripheral input so that severe disturbances of equilibrium are the exception.
- total loss of caloric response on the affected side without ever experiencing any dysequilibrium,
- slight imbalance or lightheadedness on change of head or body position, especially in the dark.
- true rotatory vertigo.

Otological Stage Imbalance

- Nystagmus may be due to either vestibular or cerebellar dysfunction.
- Mechanism of vestibular dysfunction

Otological Stage

Facial Nerve Involvement

- Although the facial nerve is compressed and maybe considerably attenuated by the expanding tumour, obvious facial weakness is uncommon. This is because motor neurons, as elsewhere in the body, are more resistant to pressure than sensory fibres.
- Facial tic
- Pain, pressure or numbness around the ear are common complaints and may be due to involvement of the sensory branch of the facial nerve.
- Nervus intermedius involvement is frequently manifested by altered lacrimation, the patient complaining of either a dry irritating eye, or of excessive tearing, and less commonly by alterations in the sensation of taste, with cachoguesia at times..

Hearing Loss

- Normal hearing
 - < 25 db HL (adults)
 - < 15 db HL (children)
- Mild hearing loss = 25-40 db HL
- Moderate hearing loss = 41-65 dB HL
- Severe hearing loss = 66-90 db HL
- Profound hearing loss = 90+db HL

Tuning Fork Tests

- The tuning fork used most commonly has a frequency of 512 Hz. The note of the higher frequency forks tends to decay quickly, not allowing sufficient time for the Rinne test to be performed. The lower frequency forks tend to enhance perception by vibration sensation

Rinne Test

- The tuning fork is struck on a bony prominence. The fork is placed firmly on the mastoid with the observer's hand steadying the head.
- The patient is asked to indicate when the sound disappears and the fork is then immediately placed erect and in line with the external auditory meatus about 2cm from the orifice.
- Positive Rinne's test – AC more than BC
- Negative Rinne's test – BC more than AC
- A conductive deafness of greater than 25 dB usually gives a negative Rinne test with a 512-Hz fork.

False Negative Rinne

- If the patient has no hearing in the test ear, the bone conduction stimulus may be perceived by the contralateral (non-test) ear, although the patient often says that he/she hears it in the test ear. As there is no hearing by air conduction, the test result is labelled Rinne negative suggesting that the deafness is conductive in nature. This mistaken impression of function in a non-functioning ear is called a false negative Rinne. In such cases the diagnosis is given by a combination of the Rinne and the Weber test. In addition, the non-test ear can be masked by a Barany noise box (a clockwork-driven sound generator of about 90dB)..

Weber Test

- The tuning fork is struck and the base placed on either the forehead, vertex or upper incisor teeth. The patient is asked where the sound is heard loudest.
- Unilateral sensorineural deafness: good ear
- Conductive deafness :affected ear.

Modified Schwabach Test

- Compares the bone conduction of the patient with the bone conduction of a normal hearing person.
- The tuning fork is placed on the patient's mastoid with the meatus blocked and, when the patient no longer hears it, the fork is placed on the normal hearing person's mastoid (usually the examiner's), again with the meatus blocked. If the examiner hears the note, the patient's bone conduction is said to be reduced.

Gelle Test

- The air pressure in the external auditory meatus is altered using a Siegle's speculum. In the normal individual, or those with a sensorineural loss, increasing the meatal pressure results in a decreased sensation of loudness from a bone-conducted stimulus. No alteration of bone-conduction thresholds indicates fixation of the stapes.
 - Bing test
- Increased loudness for bone-conducted stimuli, less than 2kHz, occurs in the normal patient or those with a sensorineural loss when the external meatus is occluded without altering meatal pressure. There is no change when a conductive deafness is present.,

Bing Test

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Clinical Tests of Balance

- ROMBERG TEST
- UNTERBERGER TEST
- GAIT TEST
- CALORIC TEST

Caloric Testing

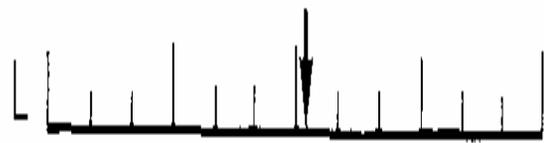
- The classical Fitzgerald—Halipike bithermal caloric test
- Supine with the head elevated to an angle of 30° to the horizontal.
- Lateral semicircular canal into the vertical plane.
- wax or perforation
- water at 44°C and 30°C (7°C above and below normal body temperature) for 40 seconds.
- Volume of water 300 ml.
- The eyes are observed for Nystagmus with the patient focusing on a near object. The end point of the nystagmus is noted and its duration recorded. Frenzel's glasses are then used to reduce visual fixation and, if the nystagmus reappears, the new end point is noted.

Caloric Test

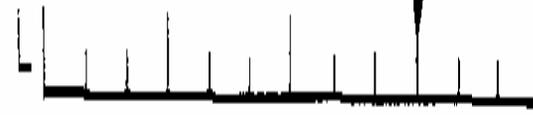
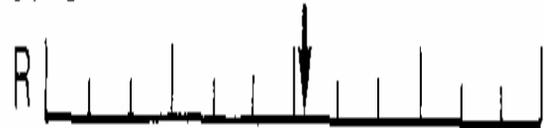
- A normal caloric reaction results in nystagmus being visible between 90 and 140 seconds after the onset of irrigation, and prolongation by a further 60 seconds following the reduction of visual fixation. The affected ear is stimulated with warm water, then the contralateral ear is tested first with warm water, then with cold, and the test concluded by cold water irrigation of the affected ear. Between each irrigation a rest period of 7 minutes is allowed.
- When the nystagmus in one direction is significantly greater after bithermal testing, it is termed 'directional preponderance' (Figure 1.9).

Caloric Test

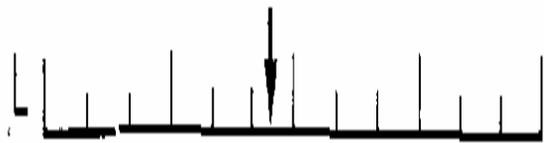
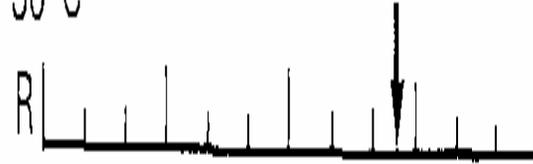
- Following bithermal caloric stimulation of a parietic labyrinth, nystagmus may be absent or decreased in amplitude and duration.
- Significant canal paresis in well over 90% of patients with an acoustic neuroma,



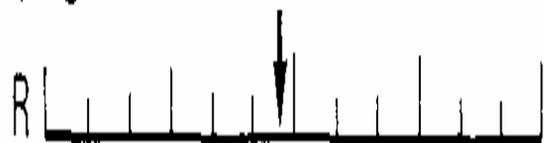
30°C



30°C

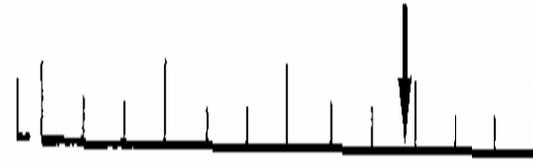


44°C

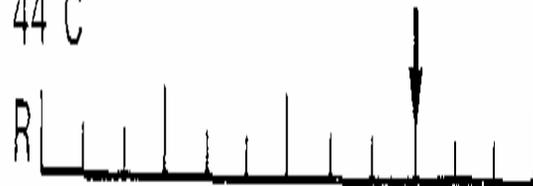


1 2 3 min

With fixation



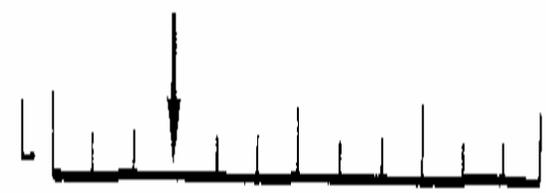
44°C



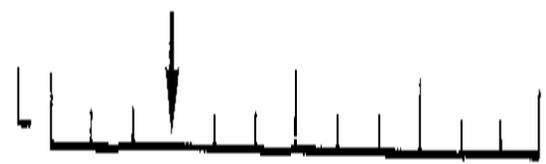
1 2 3 min

Without fixation

Figure 1.7 Caloric test: normal reaction – with and without optic fixation



30°C

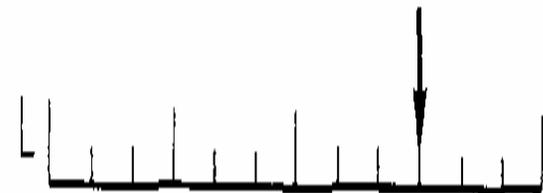


44°C

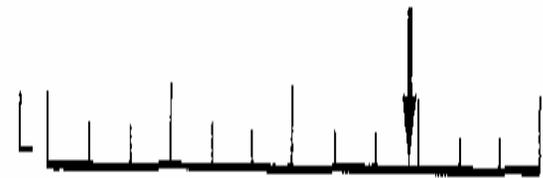
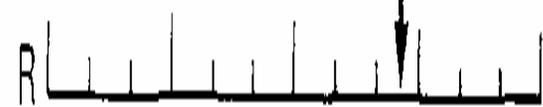


1 2 3 min

With fixation



30°C



44°C



1 2 3 min

Without fixation

Figure 1.8 Caloric test: caloric left canal paresis

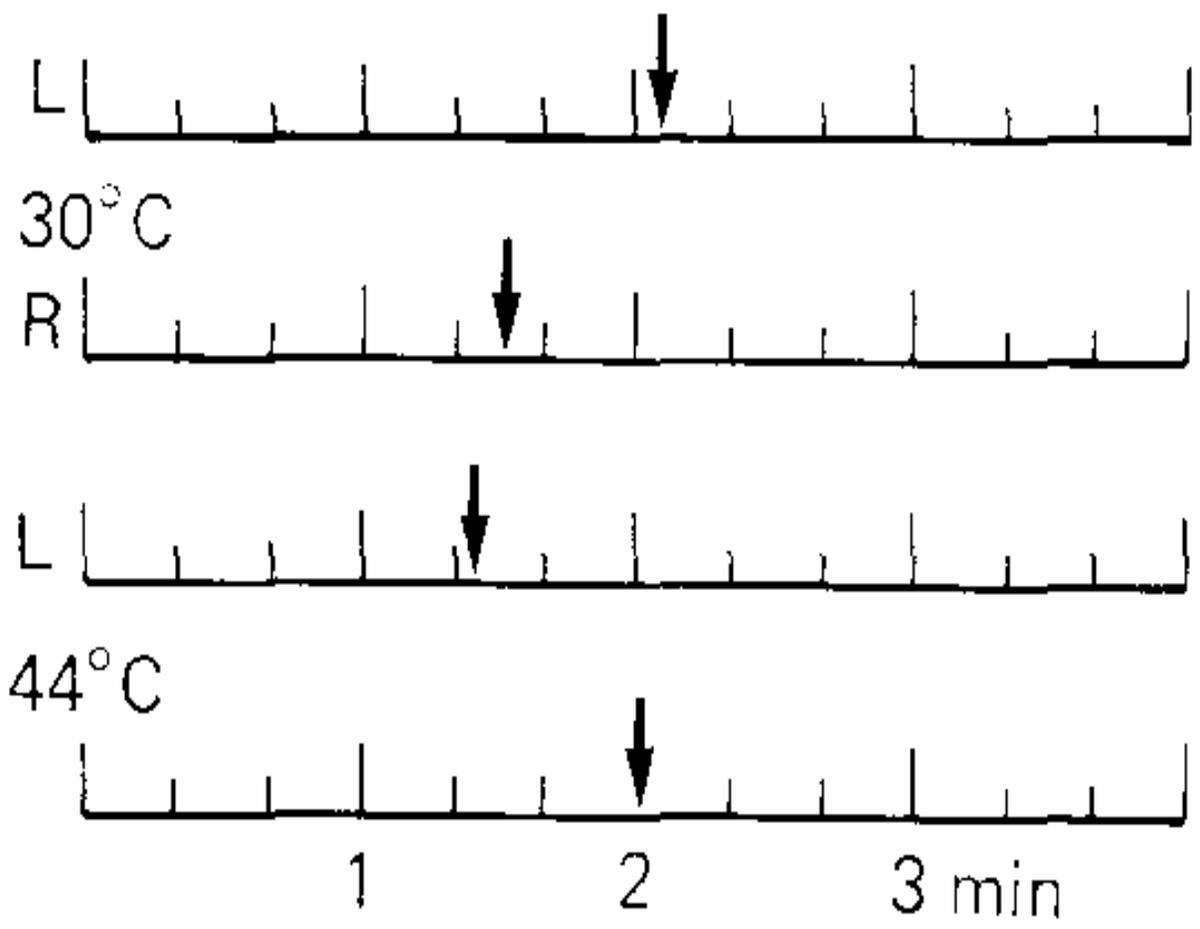


Figure 1.9 Caloric test: caloric right directional preponderance

Audiovestibular Investigation

- Pure-tone threshold
- Speech discrimination
- Stapedius reflex measurement
- Brainstem electric response audiometry
- Electrocochleography
- Loudness recruitment
- Auditory adaptation.
- Electronystagmography

Pure Tone Audiometry

- Behavioral test measure used to determine hearing sensitivity.
- Involves the peripheral and central auditory systems. indicate the softest sound audible to an individual at least 50% of the time. Hearing sensitivity is plotted on an audiogram, which is a graph displaying intensity as a function of frequency

Pure Tone Audiometry

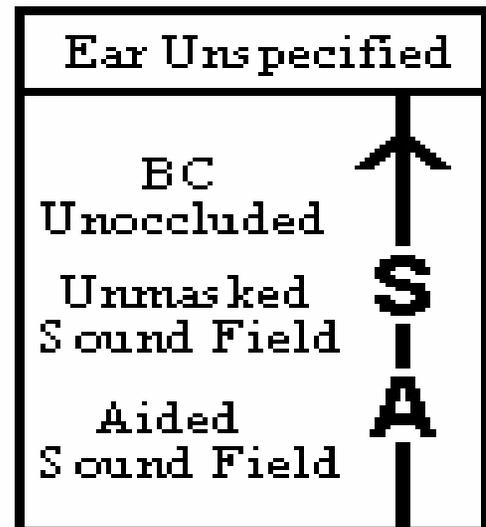
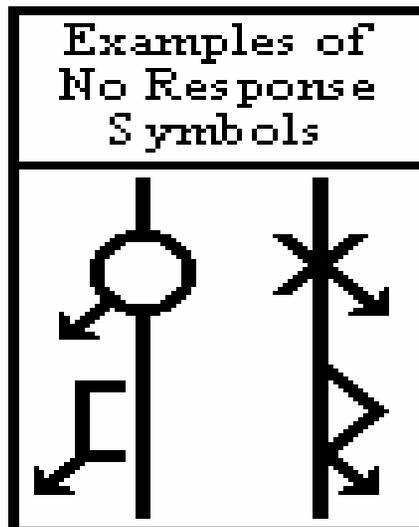
- Intensity is the level of sound power measured in decibels; loudness is the perceptual correlate of intensity.
- Frequency is cycles per unit of time. Pitch is the perceptual correlate of frequency. Frequency is measured in hertz, which are cycles per second. Usually frequencies of 250-8000 Hz are used in testing because this range represents most of the speech spectrum,.

Pure Tone Audiometry

- In pure tone audiometry, hearing is measured at frequencies varying from low pitches (250 Hz) to high pitches (8000 Hz).
- A score of 0 is normal. It is possible to have scores less than 0, which indicate better than average hearing
- Pure-tone average (PTA) is the average of pure tone hearing thresholds at 500, 1000, and 2000 Hz.

Key to Audiogram

AUDIOGRAM KEY	
Right	Left
AC Unmasked	
AC Masked	
BC Mastoid Unmasked	
BC Mastoid Masked	



TEST TECHNIQUE: Conventional

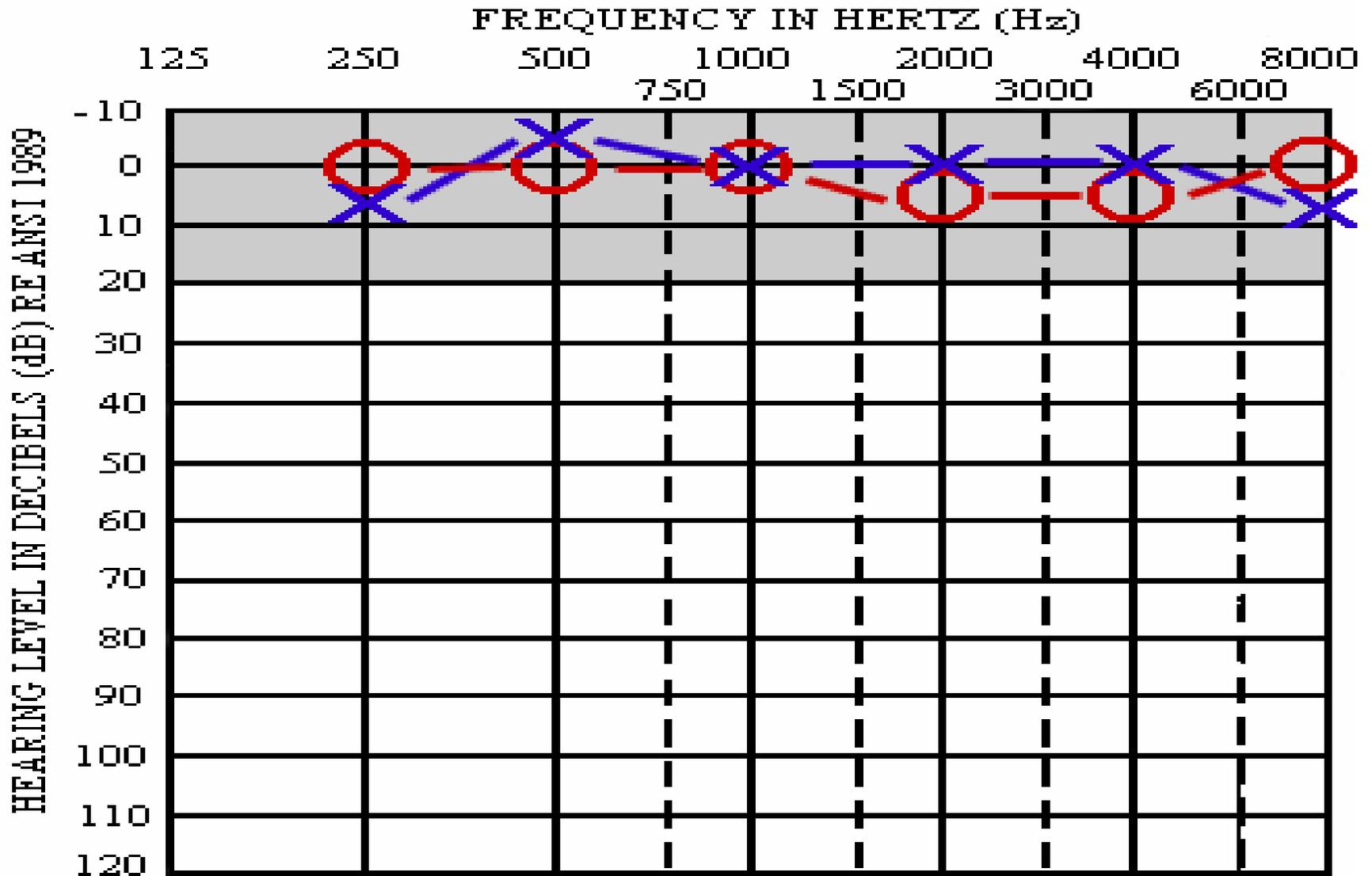
BOA VRA TROCA CPA

EARPHONE: TDH-49 TDH-50 ER3-A

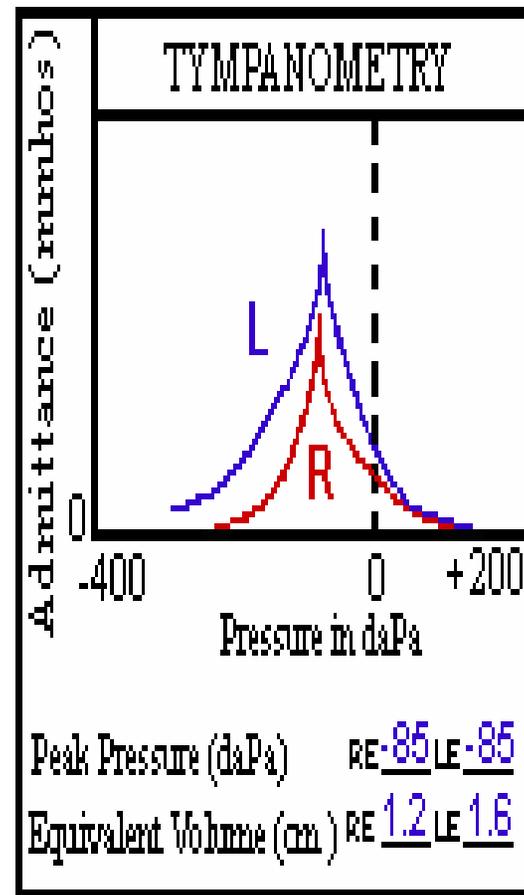
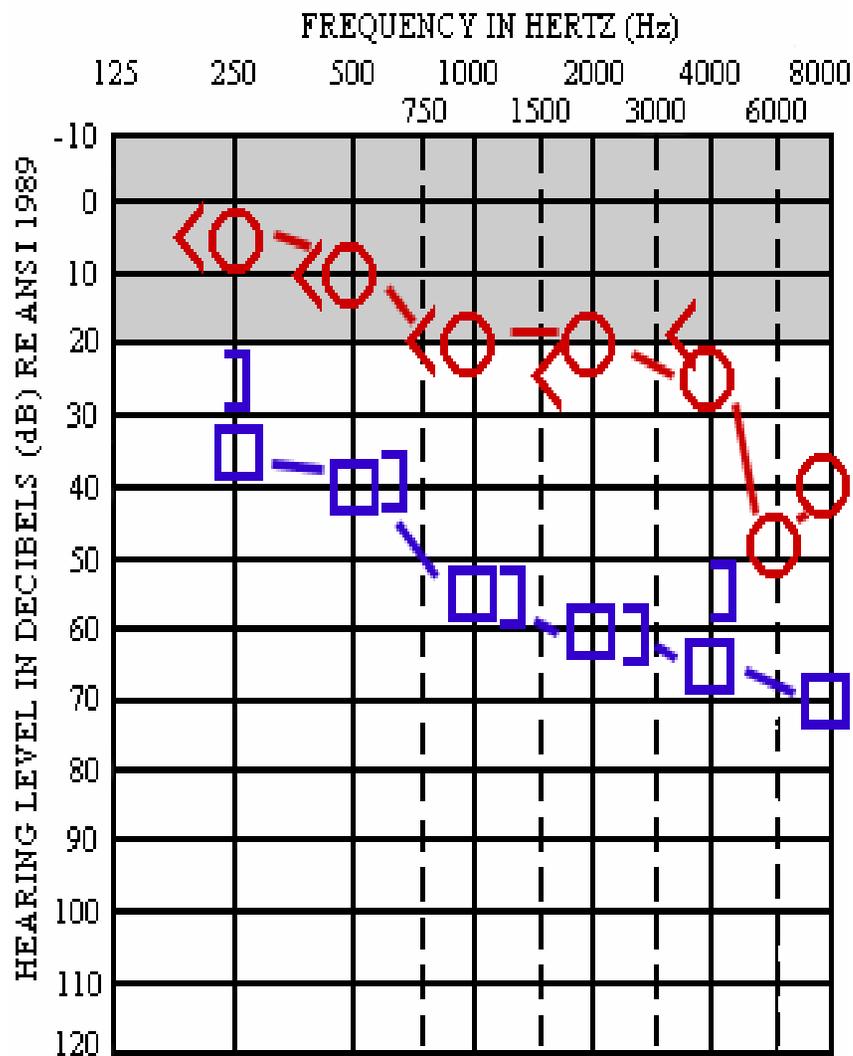
RELIABILITY: good fair poor

VALIDITY: acceptable questionable

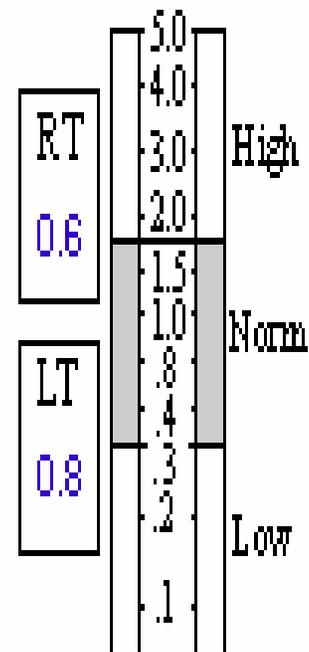
Audiogram of Normal Patient



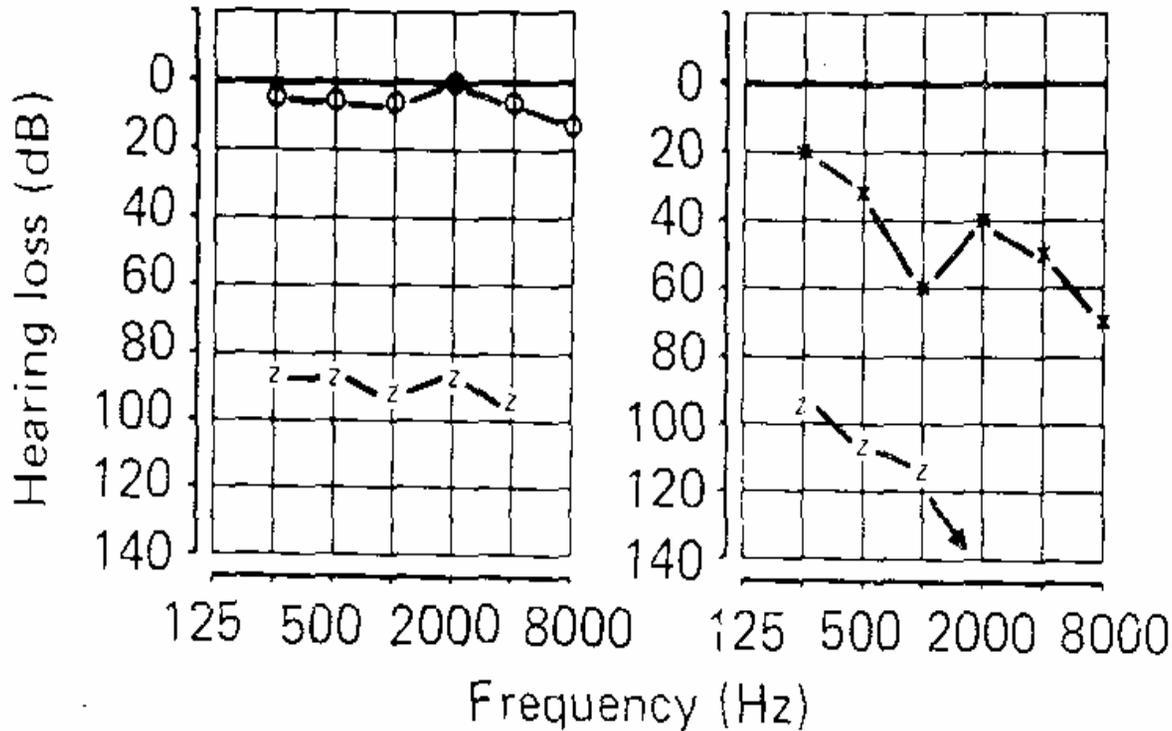
Audiogram for a Patient With an Acoustic Neuroma



Static Admittance (mmhos)
220 Hz Probe



Audiogram of a Patient With Acoustic Neuroma



Pure Tone Audiometry

- No characteristic curve for the pure-tone audiogram. Many patients have a high frequency loss, others a flat loss, some a mid-frequency notch

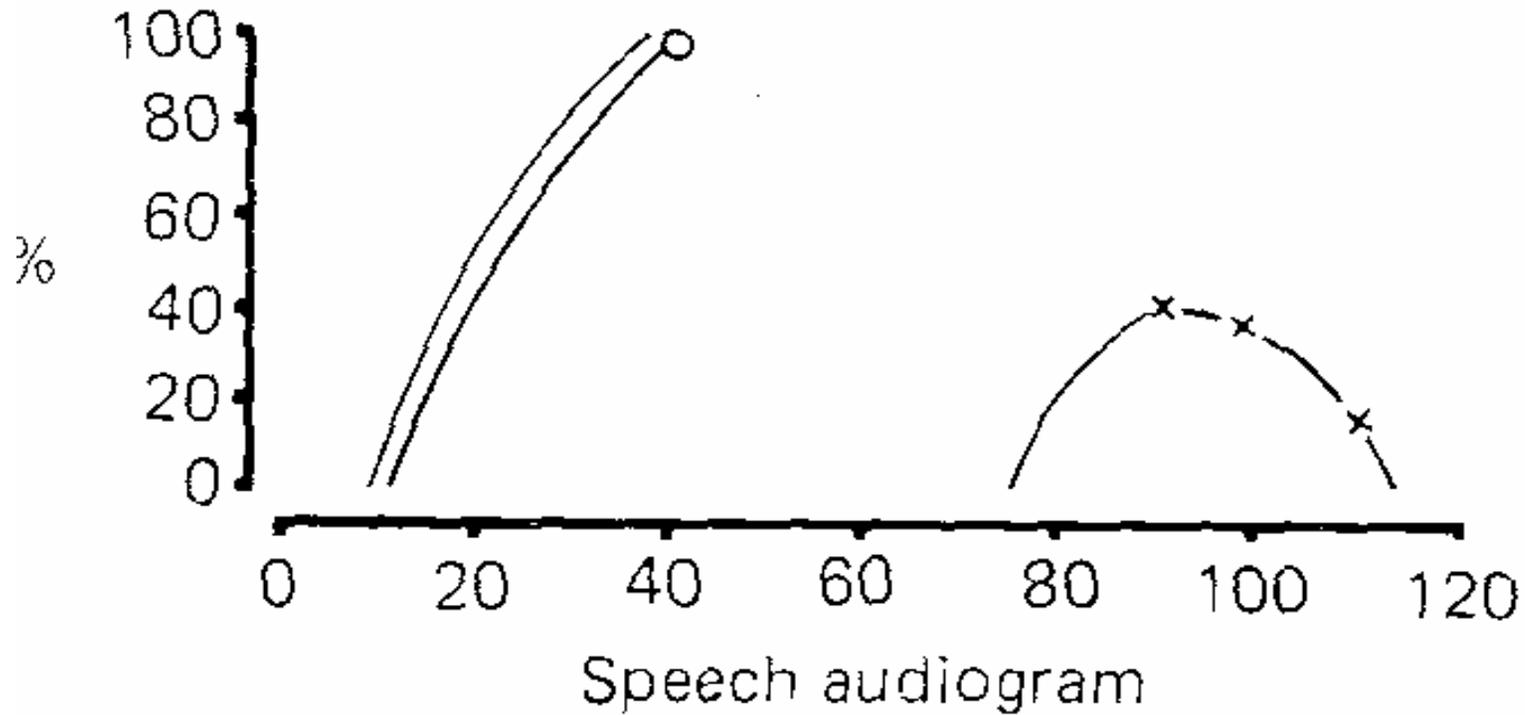
Speech Audiometry

- The cochlear nerve does not require a large population of intact neurons to transmit relatively simple pure tone messages. Speech, however basic, demands a disproportionately greater number of healthy neurons, capable of coping with the complex coding involved, particularly of temporal patterns. For this reason the typical finding in the patient with a neural lesion is of a speech discrimination score which is much worse than one would expect from consideration of the pure-tone threshold, and worse than in a patient with the same degree of cochlear deafness

Speech Audiometry

- Involves reading a list of words to see if patients can discriminate words. By comparing speech comprehension with anticipated speech comprehension, inferences can be made about central processing and central hearing deficits.
- Failure to comprehend less than 90% of the presented words is considered an abnormal result
- *Speech reception threshold*. the lowest intensity level at which the patient can correctly identify 50% of common two-syllable words such as: *baseball, airplane, mushroom..*, the pure tone average or PTA (see above) should match the SRT, within 5 dB, and the speech detection threshold (SDT), within 6-8 dB.

Speech Audiogram



Stapedius Reflex Measurement

- Retro cochlear pathology: the stapedius reflex threshold is elevated above normal levels
- Cochlear deafness: the threshold is usually normal.
- Significant elevation is 95 dB HL at 250, 500, 1000, 2000 and 3000 Hz, and 100dB HL at 1500 Hz and for the threshold to be abnormal it must be significantly raised at four out of the six test frequencies (250 Hz—3 kHz).
- Reflex asymmetry: difference in the reflex threshold between the two ears of more than 15 dB should be regarded as abnormal.
- In the majority of cases of acoustic neuroma, the elevation of the reflex threshold was greater in the higher than the lower frequencies..

Stapedius Reflex Measurement

- Stapedius reflex decay is the decline in amplitude of the reflex on prolonged stimulation, and in individuals with neural pathology the rate at which the decay occurs is increased. Pathological decay is judged to be present if the response amplitude declines by more than 50% in 5 seconds at 500 Hz and at 1 kHz.
- Abnormal stapedius reflex decay is a more specifically retro cochlear finding than elevation of the threshold.
- an elevated stapedius reflex threshold with normal stapedius reflex decay was a relatively poor indicator of the presence of a lesion in the cerebellopontine angle, whereas abnormal stapedius reflex decay with or even without elevation of the stapedius reflex threshold was highly significant.

Brainstem Electric Response Audiometry

- Within the first 7 milliseconds following acoustic stimulation, a series of five negative deflections appear). Their sites of origin are thought to be as follows:
 - N1 cochlear nerve
 - N2 cochlear nucleus
 - N3 superior olivary complex
 - N4 lateral lemniscus
 - N5 inferior colliculus

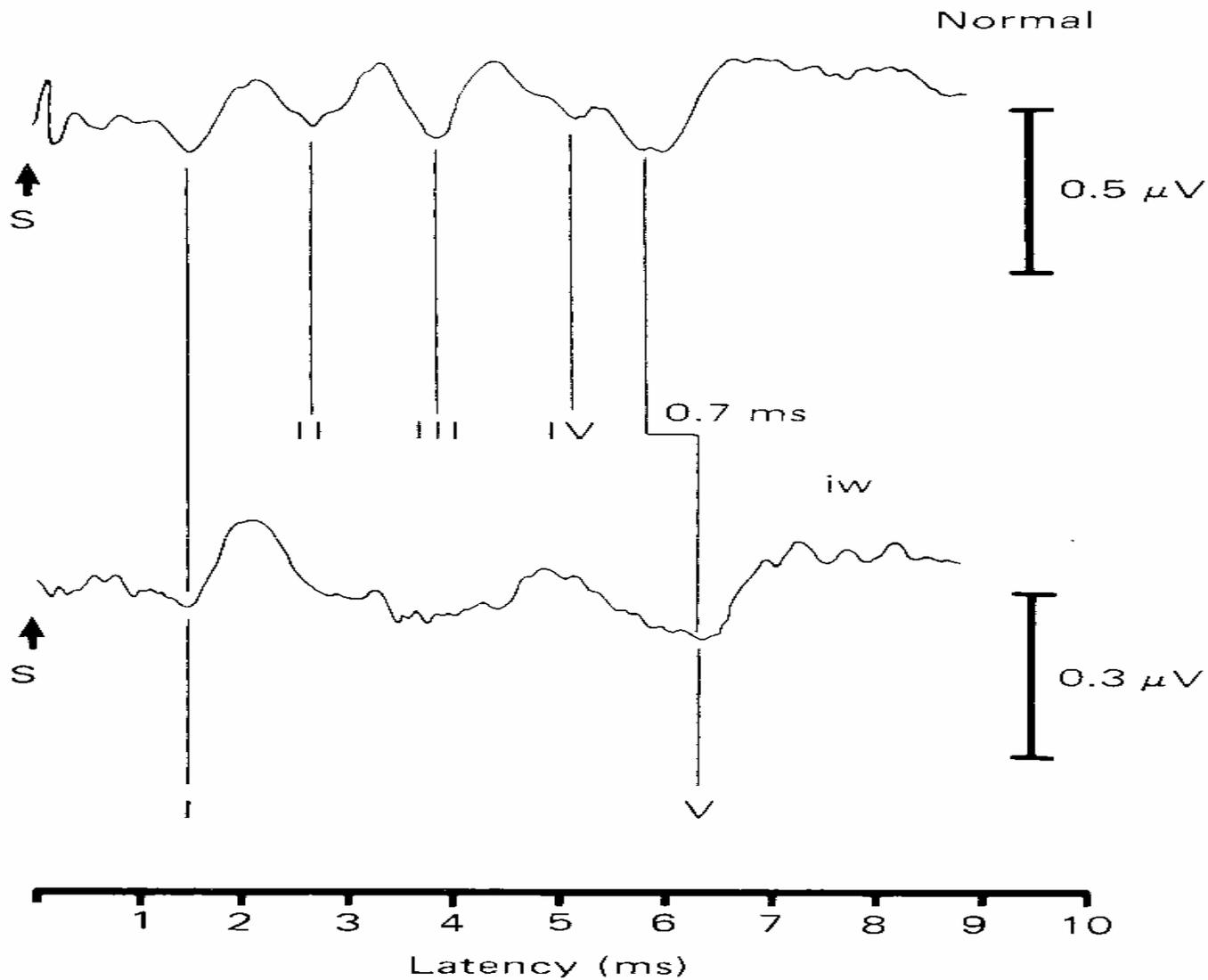


Figure 21.10 Brainstem electric response. The upper trace is from the normal ear and the lower from a case of acoustic neuroma. The latencies of N_1 are equal, but there is a highly significant 0.7 ms delay in wave V on the

Brainstem Electric Response Audiometry

- Any delay in electrical transmission in the nerve, caused for example by a tumour, would be passed on to all subsequent points in the auditory chain, and would be detectable in latency delays in wave V. which by virtue of its magnitude has proved the most convenient for study.
- Interaural latency[ITS] difference of wave V is superior to the absolute latency of wave V for the detection of acoustic tumours. Upper limit of normal for ITS, to be 0.2ms
- The chances of a subject with a normal brainstem electric response having an acoustic neuroma are very slight
- Single most reliable audio logical screening test for the condition

Brainstem Electric Response Audiometry

- The normal latency for wave V is between 5 and 5.7 ms. The interwave period between the wave 1 and wave V may be used to detect a retrocochlear lesion.
- The maximum interaural latency difference between waves 1 and V in the normal population is no more than .2 ms.

Electrocochleography

- Broadening of the eighth nerve action potential
- Good preservation of the cochlear microphonic
- Preservation of the action potential at stimulus intensities that are inaudible to the patient.

Loudness Recruitment

- Differentiation of neural from cochlear lesions. The phenomenon of 'decrement' may be seen in some cases of acoustic neuroma, that is the sensation of loudness grows more slowly in the affected ear than the normal ear.
- Recruitment, a supposed end-organ phenomenon, is 'connected with hair cell changes resulting from occlusion of the cochlear blood supply'. The procedure is, if possible, carried out at more than one frequency

Auditory adaptation

- Sound presented to the ear at a level just greater than threshold, will become inaudible after a short period of time, the length of which has a predictable value in normal ears. In ears with cochlear deafness, the values are similar to those in normal subjects, but with neural pathology, the speed of this adaptation is classically greatly increased. This phenomenon forms the basis of Carhart's tone decay test (1957).

Electronystagmography

- Examination of eye movements during several maneuvers that elicits inappropriate eye movement or nystagmus
- The most useful test in patients having suspected of having an acoustic neuroma is Barany's calorics stimulation test
- Small tumors: ipsilateral reduced response
- Large tumors: failure of fixation suppression, slowing of opticokinetic nystagmus, saccadic pursuit.