DISSERTATION ON

"COMPREHENSIVE STUDY OF AUDIOLOGICAL EVALUATION OF RETROCOCHLEAR SPACE OCCUPYING LESIONS"

Submitted in partial fulfillment of the requirements for

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of

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CERTIFICATE

This is to certify that this dissertation **"COMPREHENSIVE STUDY OF AUDIOLOGICAL EVALUATION OF RETROCOCHLEAR SPACE OCCUPYING LESIONS"** submitted by **Dr. NISHA NEELAMBARAN,** appearing for M.S ENT. Branch IV Degree examination in March 2010 is a bonafide record of work done by her under my direct guidance and supervision in partial fulfillment of regulation of the Tamil Nadu Dr.M.G.R. Medical University, Chennai. I forward this to the Tamil Nadu Dr.M.G.R. Medical University, Chennai, Tamil Nadu, India.

Director & professor,
Upgraded Institute of Otorhinolaryngolog
Madras Medical College,
Government General Hospital,
Chennai-600 003.

DEAN
Madras Medical College,
Government General Hospital,
Chennai-600 003.

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CONTENTS

INTRODUCTION

Retrocochlear space occupying lesions can be a life threatening condition presenting in different stages ranging from subtle otologic symptoms to life endangering tonsillar herniation and death.

The various retrocochlear space occupying lesions are Acoustic Schwannomas, Meningiomas, Epidermoid Cysts, Arachnoid Cysts, Schwannomas of the 5^{th}, 7^{th}, 9^{th}, 10^{th} or 11^{th} cranial nerves, Hemangiomas, Lipomas or Dermoid Cysts. Of all these, Acoustic Schwannomas in the cerebellopontine angle constitute 60- 90%. Next in frequency are Meningiomas, Epidermoid Cysts and Arachnoid Cysts.

Since these lesions are slow growing and initially may present with subtle clinical symptoms, the Otorhinologists must maintain a high index of suspicion and use appropriate audiologic , vestibular and radiographic studies to diagnose these lesions at the earliest stage. Since there is no single audiologic test to detect a retrocochlear lesion , the patient has to undergo a battery of tests which are Puretone Audiometry, Speech Audiometry, Special Tests and Brainstem Evoked Response Audiometry.

Early diagnosis if made with these tests, is of advantage since it can limit surgical morbidity and facilitate Facial Nerve and hearing preservation.and can be applied in Community Otorhinology. This study is on the efficacy of various audiological tests in the detection of retrocochlear space occupying lesions.

AIM OF STUDY

1) To study the efficacy of various audiological tests in detecting retrocochlear space occupying lesions .

2) To study the efficacy of Brainstem evoked response audiometry in screening for retrocochlear space occupying lesions .

3) To evaluate the gold standard test in the diagnosis of retrocochlear space occupying lesions.

REVIEW OF LITERATURE

Historical Perspective*:*

Vestibular Schwannomas were described in the latter part of the seventeeth century. The clinical symptom in patients with Vestibular Schwannomas were correlated with intracranial autopsy findings. By mid 1800s, neurologists understood that a tumour in the affected cerebellopontine angle patients who manifest with unilateral deafness, facial numbness and progressive blindness. The earliest attempts at removal of the cerebellopontine angle tumour was first performed by McBurney and the first successful results were achieved by Balance and Annandale. Cushing, followed by Dandy and Olivecrona initiated the next era in Vestibular Schwannoma surgery. The modern era of Vestibular Schwannoma surgery has been dominated by the efforts of House, Yasrgil, and Fisch, Glasscock, Tos and Thomsen, Palva, Sterkers, King and Morison, Samii and Wigand.

The translabyrinthine approach to the cerebellopontine angle was first suggested by Panse in 1904, first performed by Quix in 1911, and later used by Schniegelow and others to remove Vestibular Schwannomas. Benefiting from improved instrumentation and magnification , House and Hitselberger,

in the 1960s, revived the translabyrinthine approach to the lesions of the cerebellopontine angle.

Anatomy Of The Cerebellopontine Angle

The cerebellopontine angle is a triangular area bounded laterally by the medial portion of the posterior surface of the temporal bone, medially by edges of the pons and posteriorly by the anterior surface of cerebellar hemisphere and the flocculus and is part of the lateral medullary cistern. Superiorly it is limited by the Trigeminal Nerve as it crosses the petrous apex and by the edge of the tentorium. Its inferior limit is formed by the lower cranial nerves as they enter the Jugular Foramen and by the Hypoglossal Nerve. It contains an important artery, the Anterior Inferior Cerebellar Artery and two cranial nerves namely the Facial and the Vestibulocochlear Nerve as they pass from their parts of origin at the pontomedullary junction towards the Internal Acoustic Meatus.

The Internal Acoustic Meatus is a passage though the petrous bone leading to the medial wall of the vestibule. It has a porous or inlet medially and fundus laterally which is separated from the vestibule by a thin plate of bone. The lateral wall of the meatus is divide into superior and inferior

halves by the falciform crest. The upper compartment is further divided 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. The lower halve also comprises of two areas, anteriorly the Tractus Spiralis Foraminosus through which the spiraling fibres of the cochlear nerve pass and the posteriorly rather smaller area for the vestibular nerve.

Differential diagnosis of Tumours of the Internal Auditory Canal and the Cerebellopontine angle:

Although the most common tumour associated with the cerebellopontine angle syndrome is acoustic schwannoma, many other neoplastic and non-neoplastic mass lesions occur in the region.

CLASSIFICATION AND FREQUENCY OF CEREBELLOPONTINE ANGLE LESIONS:

Primary Tumours of the CPA	Revilla (1947)%	Brackmann (1980)%	Valavanis (1987)%
Acoustic Schwannoma	75.1	91.3	60.5
Meningioma	6.3	3.1	6.8
Epidermoid	6.3	2.4	3.7
Arachnoid Cyst	-	0.5	2.0
Schwannoma of the 5,7,9,10,11CN	4.9	1.4	4.0
Primary Melanoma	0.5	-	0.2
Hemangioma	-	0.3	0.7
Lipoma,Dermoid,	-	0.4	-

Thus the few most common tumours are the same in all the three series ; acoustic schwannoma, meningioma, arachnoid cyst and non-acoustic posterior fossa schwannoma. Together they account for 75% to 98% of all cerebellopontine angle lesions.

The three most common lesions in adults- acoustic schwannoma, meningioma, and epidermoid cyst – are the same in older teenagers. In younger children acoustic schwannomas are extremely rare and gliomas which are capable of enlarging the internal acoustic canal are the most common cause of cerebellopontine angle tumours[2].

ACOUSTIC SCHWANNOMAS

Acoustic schwannomas account for about 8% to 10% of all the intracranial tumours and about 60 – 90% of all the cerebellopontine angle tumours. They are schwannomas that arise from the vestibular division of the eighth cranial nerve[3,4]. Because the term acoustic neuroma is a misnomer, a subject recommended that this widely used term be replaced by the more accurate Vestibular Schwannoma. Although formerly some have maintained that superior vestibular schwannomas predominate, current opinion favours an equal origin in both superior and inferior divisions of the nerve[5].

The eighth nerve is unusual among cranial nerves in that it possesses a long segment of central myelin. It has been proposed that acoustic neuromas may arise in the region of the transition zone between the central and peripheral myelin, also known as the Obersteiner – Redlich Zone[6]. The recent literature indicates that most vestibular schwannomas originate lateral to the glial- schwannian junction of the nerve and not from the transition zone[7].

In larger tumours it is seldom possible to ascertain the precise site of origin along the nerve. Observations from smaller tumours indicate that most acoustic neuromas arise in the internal auditory canal.

MOLECULAR GENETICS:

Great studies have been made in the recent years concerning the genetic basis of acoustic neuroma formation. Through a variety of molecular genetic techniques , the gene responsible for acoustis neuroma associated with NF-2 has been localized to the long arm of the chromosome 22. The disease gene was identified in 1993, and codes for a 587 aminoacid protein. The NF-2 gene has been shown to be inactivated both by familial as well as sporadic cases. Two mutations are required to induce an acoustic neuroma- one that damages each chromosomal copy.

This" double hit" theory fits well with theories on the genetic origin of sporadic acoustic neuromas as well as those associated with NF-2. In sporadic acoustic neuromas two silencing mechanisms must occur in one cell to incite tumour growth. For this to occur it must be assumed that the gene has a high spontaneous mutation rate. Patients with NF-2 gene carry a defective copy of the gene so, when a spontaneous event inactivates the sole remaining functional copy of the acoustic neuroma suppressor gene, a tumour results.

Endocrine Relationships

Clinical series have consistently observed a slightly higher incidence of acoustic neuroma in women than men, particularly postmenopausal women[8]. Also, the onset of NF-2 may be earlier in women than men. It has also been proposed that the acoustic neuroma growth may accelerate during pregnancy. High affinity estrogen and progesterone receptors have been identified in the tumours.

Gross Pathology:

Majority of acoustic neuromas are benign, relatively slowly growing neoplasms. On gross inspection they are usually yellowish- white or gray encapsulated and frequently possess cystic components. Internally appear heterogenous and contain interspersed regions of soft and firm consistency. Their surface is typically smooth and regular. The most pronounced vascularity appears on the tumour surface, taking its origin from the numerous small vessels of the Internal Auditory canal, cerebellopontine angle, and brainstem surface.

HISTOPATHOLOGY

Histopathologic examination reveals two types of morphologic patterns; Antoni A and Antoni B. Antoni A type describes a densely packed cells with small ,spindle-shaped densely staining nuclei. Antoni B type refers to a looser cellular aggregation of vacuolated pleomorphic cells. The Antoni B morphology sems to occur predominantly in larger tumours. A whorled appearance of Antoni type A cells is a verrocay body. In Antoni B configurations the presence of large atypical hyperchromatic nuclei has been termed as ancient schwannoma by pathologists. A positive S-100 immunoperoxidase stain is confirmatory of schwann cell origin. During their

growth acoustic neuromas are “pushers” that progressively displace adjacent structures, often without macroscopic signs of invasion. Degeneration of a benign acoustic neuroma into a malignant schwannoma has been reported but is exceedingly rare[9].

Growth rate

The growth rate of acoustic neuromas has been studied both in vitro and in vivo. The fraction of dividing cells in an acoustic neuromas has been assessed by immunohistochemical means with the fraction of proliferating cells ranging between 0.36% and 3.15%[10]. When the available data are analysed acoustic neuromas increase in diameter on an average of 0.1cm to 0.2cm per year.

SYMPTOMATIC PROGRESSION OF ACOUSTIC NEUROMA WITH TUMOUR GROWTH:

Intracanalicular	Hearing loss , Tinnitus , Vertigo
Cisternal	Hearing loss , Vertigo diminishes, Disequilibrium
Brainstem compressive	Midfacial and Corneal Hypoaesthesia Occipital Headache, Ataxia Begins
Hydrocephalic	Worsening Trigeminal symptoms, Gait deteriorates, Blurred vision, Lower cranial Nerve Dysfunction , Death due to Tonsillar Herniation

CLINICAL FEATURES:

Cushing's classical description of the clinical features (described in 1917) and quoted by Shephard and Wadia :

STAGE 1	Stage of hearing disorder and vestibular manifestations
STAGE 2	Occipitofrontal Pains
STAGE 3	Incordination and instability of cerebellar origin
STAGE 4	Cranial Nerve Involvement
STAGE 5	Increased Intracranial Tension
STAGE 6	Dysarthria and Dysphagia
STAGE 7	Cerebellar crisis with respiratory difficulties

Jackler R.K has divided the growth into four anatomical stages viz.

Intracranial stage	Confined to the Internal Auditory Canal
Cisternal stage	Projects into the Cerebellopontine Angle Cistern, 7^{th} and 8^{th} nerve displaced
Brainstem Compressive Stage	Lateral surface of Pons gets compressed
Hydrocephalic stage	Fourth ventricle gets compressed 10^{th} ,11^{th}, 12^{th} cranial nerve involvement

SIGNS AND SYMPTOMS

Hearing Loss

Occuring in over 95% of patients , hearing loss is the most frequent symptom of acoustic neuromas. A number of mechanisms have been theorized. Direct eighth nerve compression , stretching of the nerve vascular compression , or occlusion of the blood supply of the eighth nerve or the cochlea , damage to the cochlear efferents , biochemical changes within the inner ear, and hemorrhage within the nerve or into the tumour all have a place in the etiology of hearing loss.

The rate and amount of hearing loss will vary depending on the rate of growth of tumours , the location within the internal auditory canal and the amount of early expansion into the cerebellopontine angle.

Schuknecht and Woellner[11] demonstrated if the Organ of Corti is intact 75% of the auditory fibres need to be destroyed before the pure tone hearing is affected. This explains the typical , slowly progressive hearing loss of cerebellopontine angle tumours. Sando[12] demonstrated anatomically that the high frequency auditory nerve fibres from the basal turn of the cochlea are located inferiorly and laterally all the way from the spiral ganglion to the cochlear nuclei in the brainstem, whereas the middle and

apical fibres twist about the axis from the spiral ganglion to the cochlear nuclei. Low frequency apical fibres are located more centrally within the nerve. These differences in the position within the nerve may be the reason for earlier involvement of high frequency basal fibres and a variable involvement of the middle and low frequency fibres from the middle and apical turns of the cochlea.

Badie and colleagues[13] proposed a theory of increased pressure in the internal auditory canal as a cause for tumour related hearing loss.

Atypical forms of hearing loss are relatively frequent. A sudden decrease in hearing occurs in some 26% of acoustic neuromas[14]. The loss may be partial or total and spontaneous recovery is possible. The diagnostic dilemma stems from the fact that only 1 -2 % of patients who have sudden hearing loss will ultimately prove to have an acoustic neuroma.

A small fraction of patients with acoustic neuromas have either normal hearing or symmetrical hearing loss. Before the era of high resolution imaging , most series showed 1-3% of patients with acoustic neuromas had normal hearing. In a recent series from the Magnetic Resonance Imaging era , 15% of patients with acoustic neuromas had subjectively normal hearing of whom 4% were audiometrically normal[15].

Tinnitus

In 70% of the patients , tinnitus accompanies the deafness in acoustic neuromas but may also present without any concomitant deafness in a small number of cases.

VERTIGO, DYSEQUILIBRIUM AND DYSMETRIA

True vertigo which is a sensation of spinning is not commonly associated with acoustic neuromas. In a recent series only 19% of individuals reported this symptom, most of whom had small tumours.
Dysequilibrium is more prevalent than vertigo, occurring in nearly 50% of patients and occurs in larger tumours[14]. Larger tumours (more than 3 cm) have a higher than 70% incidence of this symptom.

Overt cerebellar dysfunction is limited to large tumours , which indent the lateral cerebellar lobe and peduncles and impair the output of the ipsilateral cerebellar hemisphere. Truncal ataxia was found to be more common than limb ataxia.

FACIAL ANESTHESIA AND PAIN

Facial sensory disturbance occurs in approximately 50% of tumours larger than 2cm and is rarely present with smaller lesions[14]. Hypoesthesia of the midfacial region is the most common symptom. Gradually the upper and the lower divisions of the trigeminal nerve become involved. Facial Pains may also result from acoustic neuromas.

Dysfunction of the motor division of the trigeminal nerve is a symptom limited to a few advanced acoustic neuromas.

FACIAL WEAKNESS AND SPASM

The disturbances of facial mimetic function are uncommon during the natural history of an acoustic neuromas growth though the location of facial nerve is in the epicenter of the tumour growth. The facial nerve can sustain substantial compression, stretch and torsion from an acoustic neuroma and maintain its functional integrity. Facial twitching occurs in approximately 10% of patients[14], and is independent of tumour size. The most common manifestation is a minor quivering of orbicularis oculi muscle. Abnormalities in evoked electromyography and the blink reflex may be demonstrated in a minority of patients with apparently normal facial function.

The sensory component may be affected causing diminished sensation over the posterior aspect of ear canal and conchal bowl called Hitzelberger's sign which is of little practical significance.

HEADACHE

The incidence depends on the size. Very few patients with small tumours have headaches.The incidence is approximately 20% in medium sized and more than 40% in large tumours[14]. Most commonly it is either focused in the suboccipital region or generalized.

OPHTHALMOLOGIC MANIFESTATIONS

Nystagmus

A degree of spontaneous nystagmus in the horizontal plane is frequent even with small tumours. With large tumours , pronounced vertical plane nystagmus may be seen as a consequence of brainstem compression.

Brun's nystagmus is a peculiar type of gaze nystagmus found in cerebellopontine angle tumours compressing upon the pons , in which there is a large , coarse nystagmus on the side of the tumour and a fine, high frequency nystagmus on the normal side.

Decreased corneal sensation is due to involvement of ophthalmic division of trigeminal nerve and indicates a large tumour.

Papilledema from hydrocephalus is another important ophthalmologic manifestation. Hydrocephalus was found in 4% of patients[14]. Diplopia due to paralysis of fourth or sixth nerves by a large acoustic neuromas is extremely rare.

LOWER CRANIAL NERVES

Dysfunction of the lower cranial nerves ie, 9, 10, 11, 12 th is rare; making up 3.5%.

LATE SYMPTOMS

Long tract signs, depressed conciousness , and stupor, leading onto coma and respiratory arrest.

SUDDEN NEUROLOGIC DETERIORATION

Intratumoral haemorrage is a rare occurance but is a cause of sudden neurologic deterioration due to onset of an acute cerebellopontine angle

AUDIOMETRIC EVALUATION

PURETONE AUDIOMETRY

In unilateral or bilateral deafness puretone audiometry is the key investigation. High frequencies are affected first. The hearing loss may range from very mild deafness to near total deafness and sometimes normal hearing also[15].

Normal hearing is defined as :

1) Puretone average less than 20db
2) Speech discrimination score more than 90%
3) Interaural difference less than or equal to 10dB at every hertz frequency

Abnormal audiometry is defined as an interaural difference of more than or equal to 15db at a single frequency or more than or equal to 10db at two or more frequencies , and an interaural speech reception threshold difference of more or equal to 20db or a discrimination score or more or equal to 20%[16].

The pathophysiological basis for the typical hearing loss in patients with retrocochlear disease is a reduction in the number of active fibres in the acoustic nerve and it was to be expected that abnormal findings would be present especially in those tests that exert the greatest demands on the total transmission capacity of the nerve[17].

SPEECH AUDIOMETRY

In this test the patient's ability to hear and understand speech is measured. Two parameters are studied 1) Speech Reception Threshold 2) Descrimination Score

SPEECH RECEPTION THRESHOLD (SRT)

It is the minimum intensity at which 50% of the words are repeated correctly by the patient. A set of spondee (two syllable words with equal stress on each syllable) words is delivered to each ear through the headphone of the audiometer. The word lists are delivered in the form of recorded tapes or monitored voice and their intensity varied in 5dB steps till half of them are correctly heard. Normally SRT is within 10 dB of the average of puretone threshold of three speech frequencies (500dB, 1000dB, and 2000dB) .

SPEECH DISCRIMINATION SCORE (SDS)

Also called speech recognition score or word recognition score. It is a measure of patient's ability to understand speech. Here a list of phonetically balanced (single syllable words) is delivered to the patient's ear separately at 30-40 dB above his SRT and the percentage of words correctly heard by the patient is recorded. In normal persons and in patients with conductive hearing loss a high score of 90% -100% can be obtained. A score of 60-90% is found in sensorineural hearing loss, and less than 30% indicates a retrocochlear pathology.

ROLL OVER PHENOMENON

This is seen in retrocochlear hearing loss . With an increase in speech intensity above a particular level the phonetically balanced word score falls rather than maintain a plateau as in cochlear type of sensorineural hearing loss. Retrocochlear lesions usually show a roll-over ratio of more than 0.45 on speech audiometry, and have an abnormal tone decay. These two tests remain widely used and when abnormal triggers the next step in the diagnosis algorithm.

SHORT INCREMENT SENSITIVITY INDEX

Patients with cochlear lesions distinguish smaller changes in intensity of puretone better than normal persons and those with conductive or retrocochlear pathology. SISI test is thus used to differentiate a cochlear from a retrocochlear pathology.

In this test , a continuous tone is presented 20dB above the threshold and sustained for about 2 minutes. Every 5 seconds, the tone is increased by 1 dB and 20 such blips are presented. Patients indicates the blips heard. In conductive deafness , SISI score is seldom more than 15%; it is 70%- 100% in cochlear deafness and 0- 20% in nerve deafness.

THRESHOLD TONE DECAY TEST:

It is a measure of nerve fatigue and is used to detect retrocochlear lesions. Normally, a person can hear a tone for 60 seconds. In nerve fatigue , he stops hearing earlier. The threshold tone decay test is simple and is performed in the following manner.

A tone of 4000Hz is presented at 5dB above the patient's threshold of hearing , continuously for a period of 60 seconds . If the patient stops hearing earlier, intensity is increased by another 5 dB. The procedure is continued till the patient can hear continuously for 60 seconds or no level exists above the threshold where tone is audible for full 60 seconds. The result is expressed as number of dB of decay. A decay more than 25dB is diagnostic of a retrocochlear lesion.

RECRUITMENT

It is a phenomenon of abnormal growth of loudness.The ear which does not hear low intensity sound begins to hear greater intensity sounds as loud or even louder than the normal hearing ear. Recruitment is typically seen in patients with cochlear lesions and thus helps to differentiate a cochlear from a retrocochlear sensorineural hearing loss.

ALTERNATE BINAURAL LOUDNESS BALANCE TEST:

This is used to detect recruitment in unilateral cases. A tone is played alternately to the normal and affected side and the intensity in the affected ear is adjusted to match the loudness in the normal ear.The test is started at

20dB above the threshold of deaf ear and then repeated at every 20dB rise until the loudness is matched or the limits of the audiometer reached. In conductive and neural hearing loss , the initial difference is maintained throughout while in lesions of the cochlea partial, complete, or over-recruitment may be seen.

CALORIC TEST:

Because the majority of tumours arise from the divisions of the vestibular nerve, it is hardly surprising that the Bithermal Caloric Test of Fitzgerald and Hallpike (1942) frequently reveals a canal paresis. 20% of the patients with tumours of less than 2cm or less can have a normal caloric response.(Gibson and Morison 1976) Welling reported a significant canal paresis in only 66% of tumours. It does appear that in the quest for diagnosis of small tumours , the caloric test has been largely superseded by more sensitive techniques. One should remember that the caloric test produces a maximum stimulus in the horizontal semicircular canal which is innervated by the superior vestibular nerve. A small tumour arising in the inferior vestibular nerve may not cause any reduction in the caloric response.Some surgeons regard this finding as a good prognostic indication for hearing preservation, but there is no universal agreement on this point.

OTOACOUSTIC EMISSIONS (OAE)

They are low intensity sounds produced by outer hair cells of a normal cochlea and can be elicited by a very sensitive microphone placed in the external ear canal and an analysis by a computer. OAEs are present when outer hair cells are healthy and are absent when they are damaged and thus help to test the function of the cochlea. They do not disappear in eighth nerve pathology as cochlear hair cells are normal.

All patients having a unilateral sensorineural deafness should be subjected to these tests.

BRAINSTEM EVOKED RESPONSE AUDIOMETRY (BRAINSTEM AUDITORY EVOKED RESPONSE) -BERA

The auditory brainstem response is a measure of the electrical events generated in response to an auditory stimulus by components along the auditory pathway picked up by using surface electrodes.

BAER was first described by Jewett in 1970. It was first described for the diagnosis of acoustic neuromas in 1977 by Selters and Brackmann[18]. As the impulses pass through the different situations in the auditory pathway it undergoes some degree of processing at each of the stations. Passage of the

impulse through each of these pathway generates electrical activity that can be measured by placing a surface electrode on the vertex of the scalp. On graphic recording this electrical activity presents wave forms with discrete peaks the character of which is dependent upon the functional and structural integrity of the above mentioned auditory pathway. By analysis of the wave forms and its peaks as regards latency, amplitude, wave morphology, etc, we can have a fairly accurate idea about any abnormality in this pathway.

A number of responses called auditory evoked potentials can be recorded. The evoked transient responses can be recorded upto 500 milliseconds from the time of onset of the sound stimulus. Of this duration of 500 milliseconds the auditory evoked potentials of the first 10 milliseconds , called early phase of transit response , or Short Latency Response (SLR) is called the Brainstem Evoked Response Audiometry, since it records the auditory evoked potential when the auditory stimulus is traversing the brainstem region.

METHOD OF RECORDING BERA

The auditory evoked potential is elicited by a click stimulus having an intensity of approximately 45 to 65 dB above the average pure tone hearing level of the subject and is recorded with the active electrode placed over the vertex. One electrode called the reference electrode is placed on the mastoid of the ipsilateral ear and a ground electrode is placed either over the forehead just above the nasion or over the contralateral mastoid. Preferable to do a puretone audiogram prior to BERA because the sound stimulus has to be presented at a fixed suprathreshold level (usually 60dB) above the threshold and the interpretation of BERA is dependent on the audiometric contour. The sound stimulus is a broad band click of 100 microseconds duration.A broad click stimulates synchronously a large number of neurons and thereby elicites a robust BERA tracing which is clear , sharp and well demarcated with distinct and easily recognizable peaks in the graph.

The stimulus rate or click frequency is between 10 -40 clicks per second. The recording obtained is a graph plotted with amplitude in microvolts on the ordinate and time in milliseconds from the onset of stimulation on the abscissa. It consists of five to seven peaks in a normal adult appearing within 8 – 10 milliseconds.

WAVE	SITE OF NEURONAL GENERATOR
1	Cochlear nerve (distal end)
2	Cochlear nerve (proximal end)
3	Cochlear nucleus
4	Superior olivary complex
5	Lateral lemniscus ? inferior colliculus
6	Not definitely known
7	Not definitely known

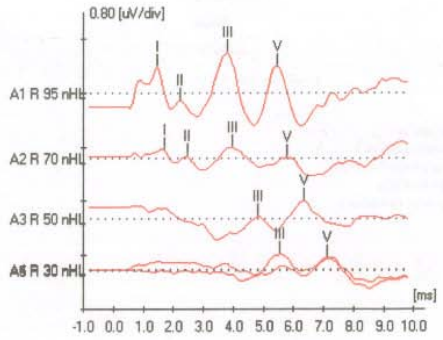
INTERPRETATION OF BERA RESPONSE

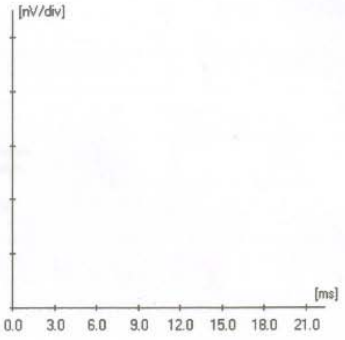
A normal BERA recording is shown in figure (i)

It has five prominent peaks and two small peaks , which are named from 1 to 7 , each giving information about a specific segment of the auditory pathway from cochlea to brainstem; hence the need of identifying each wave accurately.

Wave 5 is the most reliable and easily identifiable wave in BERA tracing. The hallmark of wave 5 is that there is a sharp negative deflection immediately following a peak beyond 5 milliseconds mark.

NORMAL BERA WAVES





Latencies (ms)

Label Index	*I*	*II*	*III*	*IV*	*V*
A1	**1.45**	**2.24**	**3.82**		**5.45**
A2	**1.70**	**2.49**	**3.99**		**5.78**
A3			**4.82**		**6.36**
A5			**5.53**		**7.11**

Interlatencies (ms)

Label Index	*I-III*	*III-V*	*I-V*
A1	**2.37**	**1.62**	**4.00**
A2	**2.29**	**1.79**	**4.08**
A3		**1.54**	
A5		**1.58**	

Stimulus Parameters

Label Index	*Intensity*	*Ear*	*Transducer*	*Insert Delay*	*Type*	*Frequency*	*Polarity*	*Ramp*	*Rise/Fall Plateau*	*Rate*

Wave 4 is identified as the peak just preceding wave 5. A distinct and separately identifiable wave 4 is present in only 50-60% of subjects. In other cases the wave cannot be identified separately.

Wave 3 is identified as the upward peak between Wave 2 and Wave 4, just beyond the 3 millisecond on the graph. It is consistent as Wave 1 and Wave 5 and may be absent in only about 1-2% of cases. It is normally present around the 3.8 millisecond mark on the BERA graph.

Wave 2 is the peak immediately preceding the easily recognizable Wave 3 and has a latency of approximately 2.8 milliseconds.

Wave 1 is the sharp peak just beyond the 1 millisecond mark on the BERA graph, usually present at 1.65 to 1.75 milliseconds. It is a consistent wave and is present in all normal subjects.

The parameters used frequently are :

1) Ipsilateral latency of potential Wave 5 as compared to normal.
2) Difference of latency of potential Wave 5 of ipsilateral and contraleral sides.
3) Ipsilateral latency difference of Wave 1 to 5 as compared to normal.

4) Difference of latency potentials of Waves 1 and 5 of ipsilateral and contralateral side.

Comparing latency difference of potentials of ipsilateral and contralateral sides was the safest method to avoid a false positive diagnosis[19].

The absolute latency of Wave 5 is most important for clinical measurement because it is commonly present and easily identifiable and because the latency of Wave 5 is dependant upon the intensity of sound stimulus.

The interaural latency of a particular wave should not be more than 0.2 millisecond provided the same suprathreshold sound is presented to both ears . If the difference is more than 0.3 milliseconds existence of some lesion in the vestibular pathway should be suspected. The interwave latency measured shows any lesion between the cochlear nerve and the brainstem. It is approximately 4 milliseconds between Waves1 and 5.

AMPLITUDE STUDIES

Amplitude studies are not a very good criteria since the amplitude of the waves is not as constant as latency of the waves.They are variable and so the clinical information obtained are not much reliable.

WAVE MORPHOLOGY

Wave morphology ie, the shape or configuration of the graph that is obtained on evoked response audiometry also provides some clinical information. Any retrocochlear lesion may alter the morphology of the waves.

BERA is the most sensitive and specific test in the audiology battery for detecting disorders that affect the brainstem. It is an inexpensive and noninvasive monitor of brainstem status in patients with confirmed or suspected brainstem disorders[20].

SENSITIVITY OF BERA IN DETECTING ACOUSTIC NEUROMAS IN VARIOUS STUDIES[21]

The overall sensitivity is 90%

Roland(1987)	94%
Harder(1988)	100%
Josey(1988)	97%
Moffet(1989)	93%
Welling(1990)	92%
Grabel(1991)	91%
Wilson(1993)	85%
Dornhoffer(1994)	93%
Thomson(1994)	98%

RELATIONSHIP BETWEEN TUMOUR SIZE AND BERA SENSITIVITY

Study	**Tumour Size**	**Sensitivity**
Hashimoto et al	<1.5 cm	78%
Wilson et al	<1.5 cm	85%
Gordon & Cohen et al	<1 cm	69%
Zappia et al	<1 cm	89%
Dornhoffer et al	<1 cm	93%

THE STACKED ACOUSTIC BRAINSTEM RESPONSE

The stacked Acoustic Brainstem Response was proposed by Don et al in 1997 and reflects the loss of synchronized neural fibre activity, no matter which fibres are compromised by the tumour. It is sensitive to neural fibre activity from all frequency regions of the cochlea.

A click stimulus is used to activate the whole cochlea and the resulting response is separated into five frequency bands and are called derived band Acoustic Brain Responses and are used in constructing the stacked ABR.

In a series of 25 tumour cases, Don et al (1997) found that five small (<1cm) intracanalicular tumours were missed by standard ABR latency measures. However they demonstrated that all were detected by this new stacked ABR method.

The main reason for the sensitivity of stacked ABR is that the temporal alignment and summation of derived waveforms to form a stacked ABR generate a stacked amplitude that reflects the total synchronous neural response to the stimulus. Thus the activity of all activity of neural elements contribute to the stacked amplitude. Elimination of any significant amount of

synchronous neural activity by the tumour will result in a significant reduction in the stacked ABR amplitude. Thus it is more sensitive than the standard latency and amplitude measures that require compromise of specific neural elements.

A major limitation of stacked ABR is that it must be obtained with click levels no greater than 60-65 dB nHL, because masking noise greater than this can be uncomfortable.

IMAGING MODALITIES

There have been four major eras in the evolution of imaging technology in the diagnosis of acoustic neuromas.Plain films of the internal acoustic meatus (1910s), Polytomography (1950s) Computed Tomography(1970s), Magnetic Resonance Imaging (1980s). Both plain films and polytomography rely on detection of osseous changes involving Internal Auditory Canal, and so were not sensitive . By comparing the normal Internal Auditory Canal to the abnormal side and using the criteria of a difference in canal height of more than 2mm, a shortening of the posterior wall of the canal of more than 3mm , a downward displacement of the crista falciformis , and the presence of focal erosion, Valvassori found abnormal Internal Auditory Canals on polytomography in 78% of surgically verified

tumours[22]. Contrast enhanced Computed Tomography and gas contrasted Compued Tomography improved the sensitivity but still small lesions were overlooked.

The introduction of Magnetic Resonance Imaging in the early 1980s was a major improvement in acoustic neuromas diagnosis .Gadolinium enhanced Magnetic Resonance Imaging further increased the sensitivity and is still the gold standard investigation[23].

Acoustic Schwannomas range in size from a few millimeters to 6 cm-8cm large tumours. A small percentage of tumours are entirely intracanalicular , majority are medium sized schwannomas (upto 2 cm) centered at the acoustic porus and a stem of tumour that extends into the Internal Auditory Canal, appearing as an ice-cream cone or mushroom. The very large tumours arise from the cisternal portion of the nerve and have little intracanalicular component.

On non-contrast enhanced Computed Tomography 64% of acoustic neuromas are isodense. Nearly 90% of the untreated tumours enhance homogenously with contrast.

On Magnetic Resonance Imaging , medium sized tumours are most likely to be homogenous , and large tumours are likely to contain internal zones of inhomogenity.

On T1-weighted images , schwannomas are usually isointense or mildly hypointense relative to the pons and hyperintense to CSF. On T2-weighted images they are mildly hyperintense to the pons and iso- to hypointense to CSF. Thus CSF becomes a natural contrast for detecting the tumour.

Acoustic schwannomas enhance intensity after intravenous administration of a gadonilium contrast agent on T1-weighted images. The sensitivity of contrast – enhanced, T1-weighted Magnetic Resonance Imaging can approach 100% in detecting schwannomas[21,24]. In a recent series , both contrast T1-weighted and high resolution T2-weighted images detected tumours ranging from 0.06 to 3.0 cm[22,23].

MANAGEMENT

Three treatment options are currently available:

1) Observation with serial imaging
2) Microsurgery
3) Stereotactic radiation

The choice of option is based on the preservation of life considering the natural course of these benign tumours and possible neurologic sequelae(eg, cranial nerve dysfunction, ataxia), tumour removal, preservation of facial nerve function , and preservation of hearing.

Conservative management is recommended for patients with small tumours who have a good possibility of not needing any treatment in their predicted natural life span. The obvious advantage of “wait and scan” is that it avoids a potentially morbid intervention.

MICROSURGICAL MANAGEMENT

The priorities in acoustic neuromas surgery are the preservation of life, the maintainance of facial nerve function and the preservation of socially useful hearing in the tumour ear. Surgical procedures take place in two broad stages (1)Craniotomy and exposure of the tumour

(2)Microdissection of the tumour away from brain, cranial nerves and adjacent vascular elements. The most commonly used procedures are the retrosigmoid , translabyrinthine and middle fossa approach, others are transotic and extended middle fossa technique.

STEREOTACTIC IRRADIATION:

A precise , conformal dose of radiation with isodose lines tailored to the center margins of the tumour is given. A single large dose of irradiation is given in a single session to induce necrosis in the tissue , approximately 25 Gy to the center and 12-14 Gy to the margins[25].

MENINGIOMAS

Meningiomas arise from the meningothelial arachnoid cell and account for 13% - 18% of all the primary intracranial tumours. In the cerebellopontine angle it is the second most common and is a difficult differential diagnosis.

They are broad based against a dural surface such as the posterior petrous wall and are usually eccentric to the porous, unlike acoustic schwannomas. Meningiomas frequently (56%) herniated into the middle fossa and are moderate to large tumours.

Meningiomas are either isodense (31%) or hyperdense (69%) on non-contrast Computed Tomography and hypointense or isointense on T1-weighted images but are variable in intensity on T2-weighted images on Gadolinium Magnetic Resonance Imaging dural thickening surrounding a meningioma (“ dural tail, a meningeal sign or flair sign “) is seen in 52-72% of cases. Dural thickening bordering a meningioma extending into the Internal Auditory Canal may simulate the stem of an acoustic neuroma. The treatment of a meningioma is surgical. Local recurrences are not uncommon.

EPIDERMOID AND OTHER CYSTS

Also called primary cholesteatoma, or pearly tumours, congenital intradural epidermoid cysts are the third most common tumour in the cerebellopontine angle. They present in middle adulthood, though congenital they tend to expand where physical resistance is low and so have variable shapes. On Computed Tomography they are nonenhancing and approximate

CSF in density.On Magnetic Resonance Imaging epidermoid cysts tend to be relatively homogenous, and are usually isointense or slightly hyperintense to CSF on T1- weighted images and are hyperintense to CSF on T2 weighted images.

ARACHNOID CYST

They are usually large masses, possess smoother surfaces than epidermoids and on Computed Tomograms, intrathecal contrast may be used to differentiate their surface characteristics. They are of the same density and intensity of CSF and are difficult to differentiate. Their symptoms may be controlled by diuretics.

Cysticercosis is another diagnostic consideration in endemic areas. Lipomas and Dermoid cysts of the Internal Auditory canal and Cerebellopontine Angle can also cause Retrocochlear hearing loss.

MATERIAL AND METHODS

Patients with diagnosed cerebellopontine angle space occupying lesions who came to the Neurotology Department of Upgraded Institute of Otorhinolaryngology of Government General Hospital, Chennai between the years 2007 – 2009 were included in the study, which consists of 30 cases.

Inclusion Criteria:

1) Patients radiologically proven to have retrocochlear space occupying lesions.
2) Patients having otologic symptoms.
3) Age group above 15 years .
4) Patients willing for the study.

Exclusion Criteria:

1) Patients with no otologic symptoms.
2) Patients less than 15 years.
3) Patients with life threatening complications.
4) Patients not willing for the study.

A total of 30 patients with radiological proven retrocochlear space occupying lesions and with otologic symptoms were evaluated audiologically, with Puretone Audiogram, Speech Audiometry, Caloric tests, and Brainstem Evoked Response Audiometry using a standardized case record form and statistics evaluated and compared against standard series for reference.

TEST METHODS

PURETONE AUDIOMETRY

Puretone audiometry was done using MAICO MA52 audiometer. (Fig ii) The method used was modified Hughson Westlake ascending and descending method. Starting at 70dB reduce 10dB till the patient stops hearing. Then increase 5 dB each time till the threshold ie, minimum intensity, at which the patient can hear is reached .

Hearing Loss is Graded as Follows:

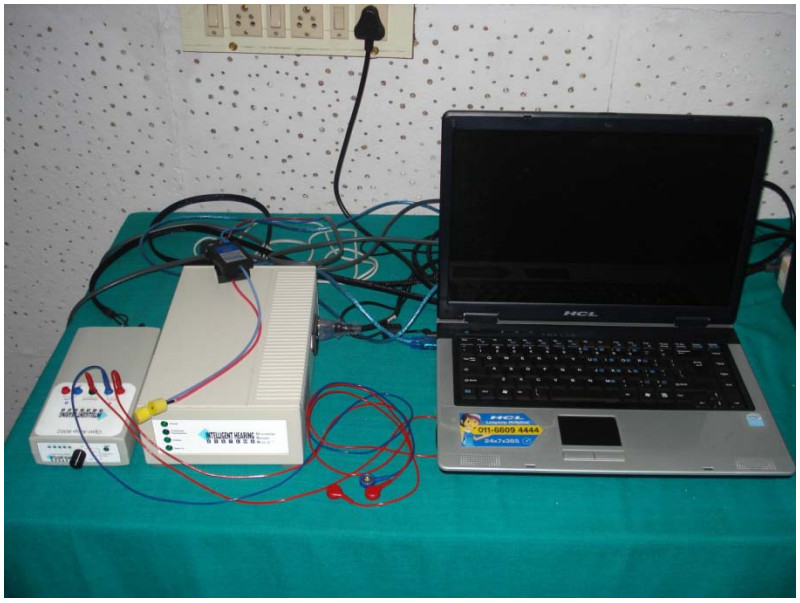
0 - 25dB	Normal
25- 40dB	Mild
40- 55dB	Moderate
56- 70dB	Severe
71- 90dB	Profound
>91dB	Total Hearing Loss

SPEECH AUDIOMETRY

The audiometer used was MAICO MA52.

PURETONE AUDIOMETER

BERA MACHINE



Speech Reception Threshold

The test begins with 25dB above the puretone threshold. A test of 10 “spondee” words are the stimuli 25dB above the puretone average, which is reduced by 10dB till no response is obtained. Then increased by 5dB till the patient identifies 50% of the words.Repeat the procedure. The minimum intensity level at which the patient is capable of repeating 50% of the speech stimuli is the speech reception threshold.

Speech Descrimination Score

Phonetically balanced words are presented 40dB above the Speech Reception Threshold. A list of 50 words are given to the test ear and the percentage of correct responses is calculated. A score less than 30% indicates a retrocochlear pathology.

SPECIAL TESTS

1) Tone Decay Test

Olsen and Noffsinger method was used. 20dB suprathreshold stimulus is given to the test ear continuously for 1 min. If the patient hears for 1 minute the test is negative and if it stops before 1 minute, it is positive and indicates retrocochlear pathology. This is done with 500Hz to 4kHz frequency.

2) Suprathreshold Adaptation Test

White noise of 90dB at 1kHz is used to mask the normal ear and 110dB SPL is presented to the test ear continuously for one complete minute. If the patient hears for one complete minute the test is negative . If the patient hears for less than a minute the test is positive for a retrocochlear pathology.

3) Short Increment Sensitivity Test

The test begins at 20dB above the puretone threshold and at a frequency of 1kHz. 1dB increment is presented to the test ear twenty times. The percentage of the identified increments are calculated. A score of more than 70% is indicates a cochlear pathology.

4) Alternate Binaural Loudness Balance Test

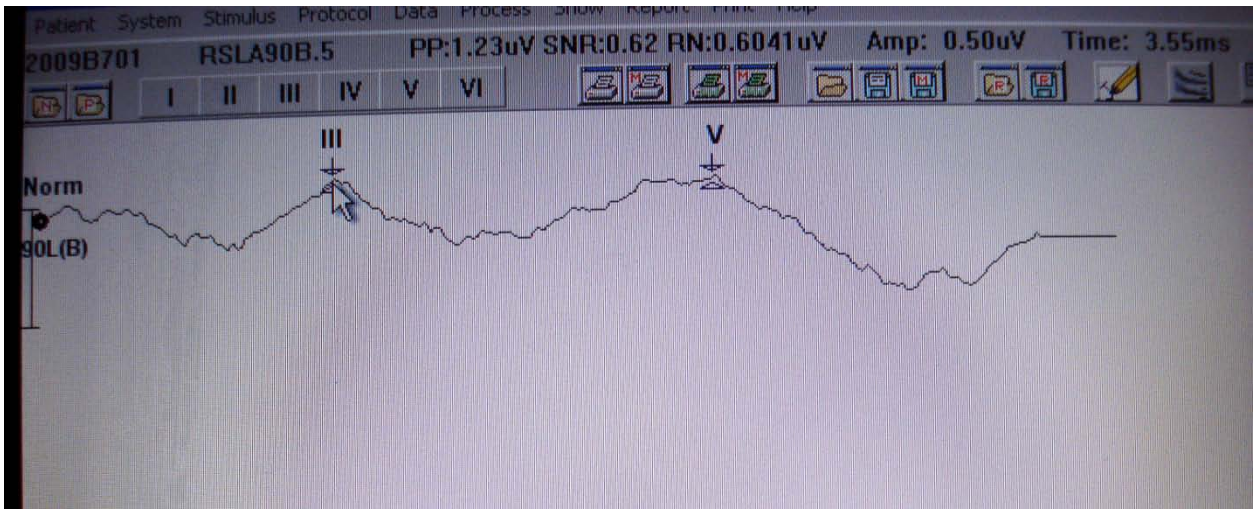
From the threshold, 10dB is increased each time and the increment in intensity is compared with the opposite ear and plotted on a laddergram, and inference arrived based on the presence of recruitment in cochlear pathology.

Brainstem Evoked Response Audiometry

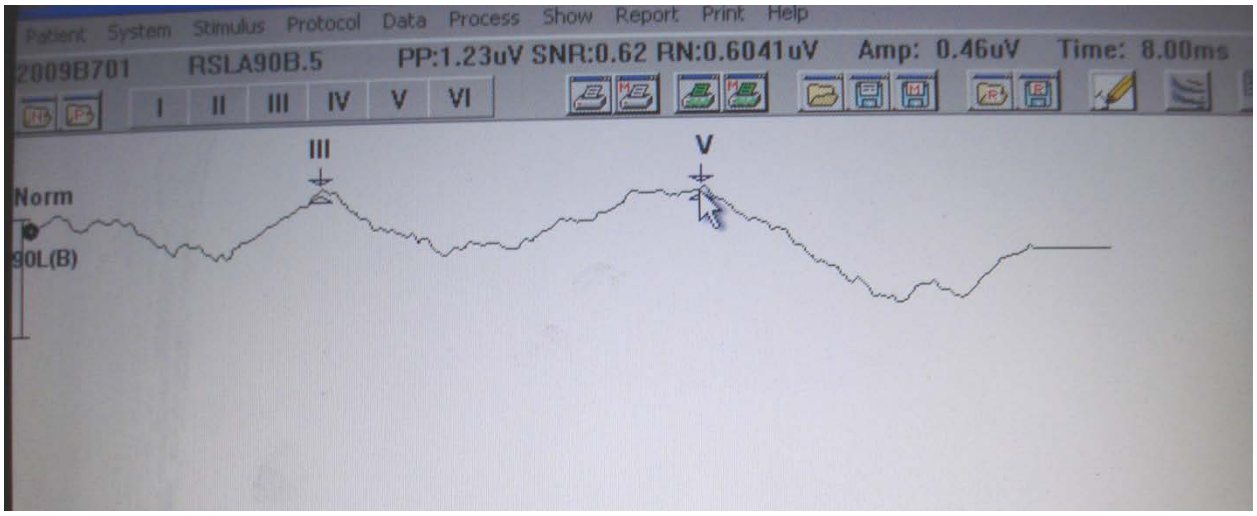
BERA was performed using USB Jr. (Universal Smart Box Jr.); Intelligent Hearing System. (Fig ii) Testing was conducted in a sound attenuating room with the patient in supine position.Scalp electrodes were placed at the vertex , high forehead and over the mastoids .A dual channel recording was thus obtained. The impedence was kept below 5000ohms. Rarefaction click stimuli at 90dB nHL at a rate of 11.1.per second were delivered through TDH-39 aural earphones.The band pass fillers were set at 150-3000Hz (12dB per octave rolloff).The analysis time was set at 10ms and 1024 average obtained for each recording. Atleast two trials performed on each side to get reproducible recordings. The threshold for intreaural latency difference was set at 0.3 ms.

BERA WAVES IN RETROCOCHLEAR LESIONS

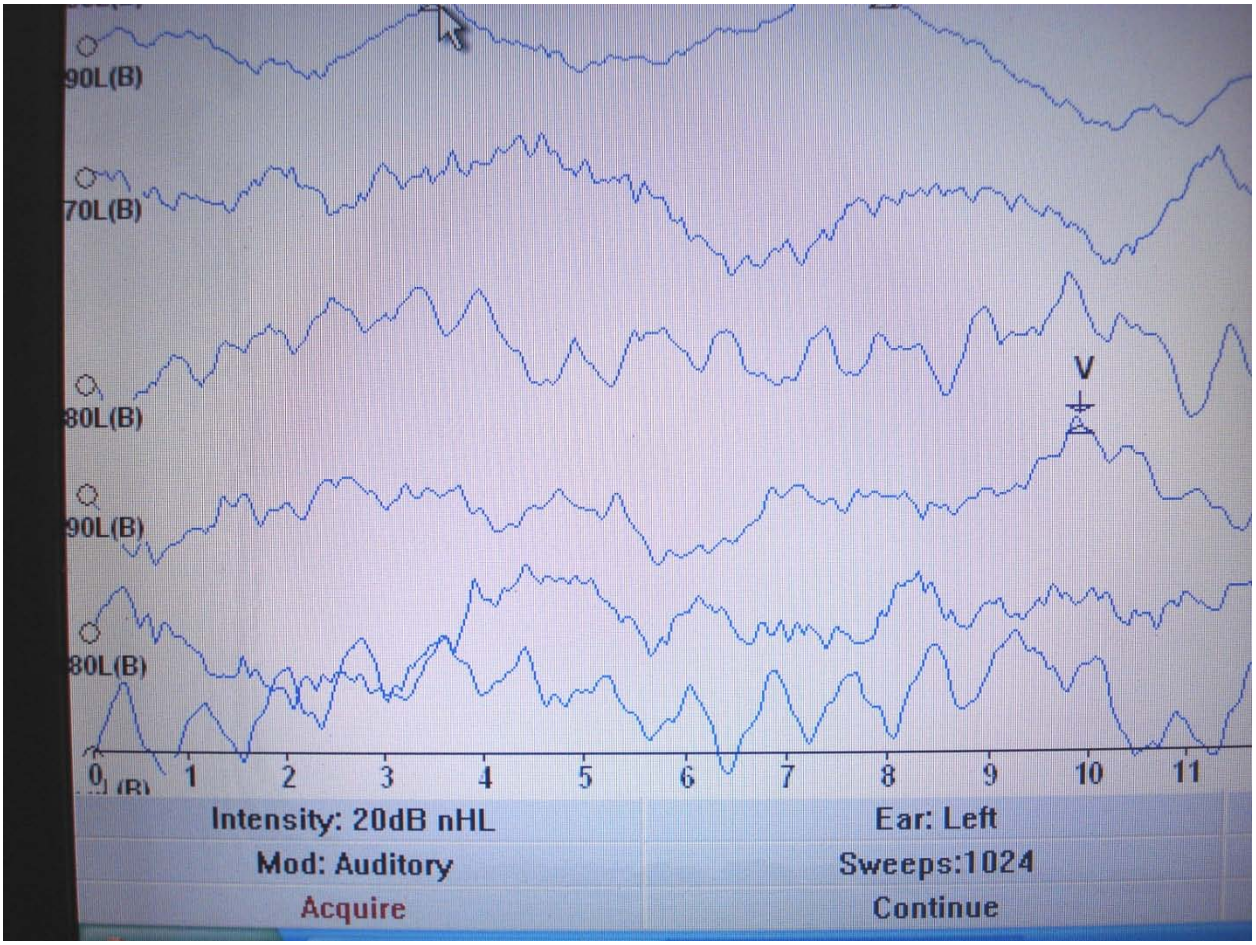
WAVE III at 90 L(B) shows normal absolute latency



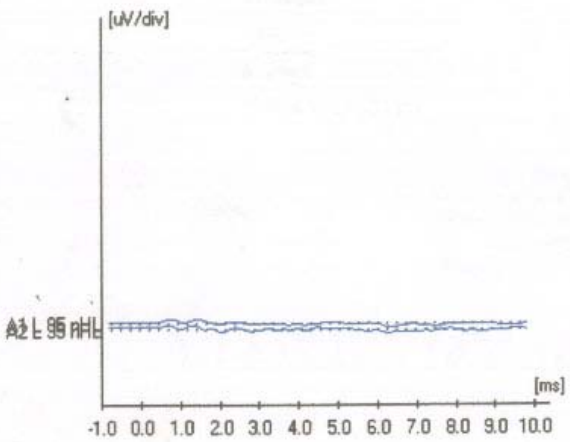
Wave V at 90 L(B) shows increased absolute latency (8.00ms)

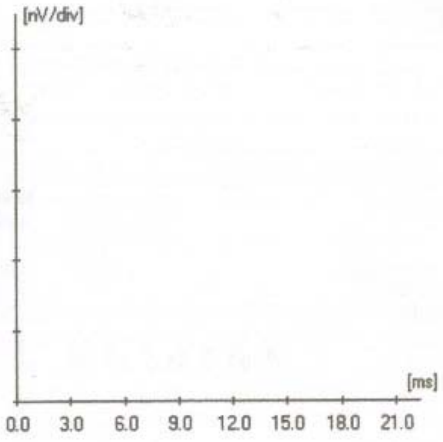


BERA WAVES SHOWING INCREASED ABSOLUTE LATENCY FOR WAVE V AND POOR MORPHOLOGY



BERA REPORT SHOWING NO REPONSE AT 95 nHL





Latencies (ms)

Label Index	*I*	*II*	*III*	*IV*	*V*

Interlatencies (ms)

Label Index	*I-III*	*III-V*	*I-V*

Stimulus Parameters

Label Index	*Intensity*	*Ear*	*Transducer*	*Insert Delay*	*Type*	*Frequency*	*Polarity*	*Ramp*	*Rise/Fall*	*Plateau*	*Rate*
A1	**95dB nHL**	**Left**	**Insert Earphones**	**0.80**	**Click**	**N/A**	**Rarefaction**	**N/A**	**N/A**	**N/A**	**31.11**
A2	**95dB nHL**	**Left**	**Insert Earphones**	**0.80**	**Click**	**N/A**	**Rarefaction**	**N/A**	**N/A**	**N/A**	**31.11**

Recording Parameters

Label Index	*Epoch*	*Points*	*Pre/Post*	*Averages*	*Artifacts*
A1	**10.66**	**256**	**0.00**	**2000**	**3**
A2	**10.66**	**256**	**0.00**	**2000**	**1**

Criteria Used for Diagnosis of Retrocochlear Pathology (Fig iii&iv)

1) Prolongation of absolute latency of waves 1 , 3 , 5
2) Prolongation of interaural latency 1-3 , 3-5 , or 1-5
3) Poor wave form morphology
4) Absent wave forms

Magnetic Resonance Imaging

Axial , Coronal , Sagittal sections with T1 weighted and T2 weighted images were done.Gadonilium- DTPA was given at a dose of 0.1 mmol/kg to look for enhancement of T1 weighted Magnetic Resonance images by its selective concentration in the tumours. The contrast material enters these tumours because of absence of blood-tissue barrier in the vascular bed of the tumour. These images were studied with special reference to the internal auditory canal and cerebellopontine angle to detect the abnormalities.

RESULTS AND ANALYSIS

DEMOGRAPHIC PROFILE OF THE STUDY AGE (Fig. 1)

The age of cases ranged from 18 to 72 years.

The mean age was 43.4 years.

The median age was 44.5 years.

The mean age of acoustic schwannoma was 40.7 years.

The mean age of meningioma was 58.8 years.

SEX (Fig. 2)

Females were found to be more affected.

Males - 46.7%

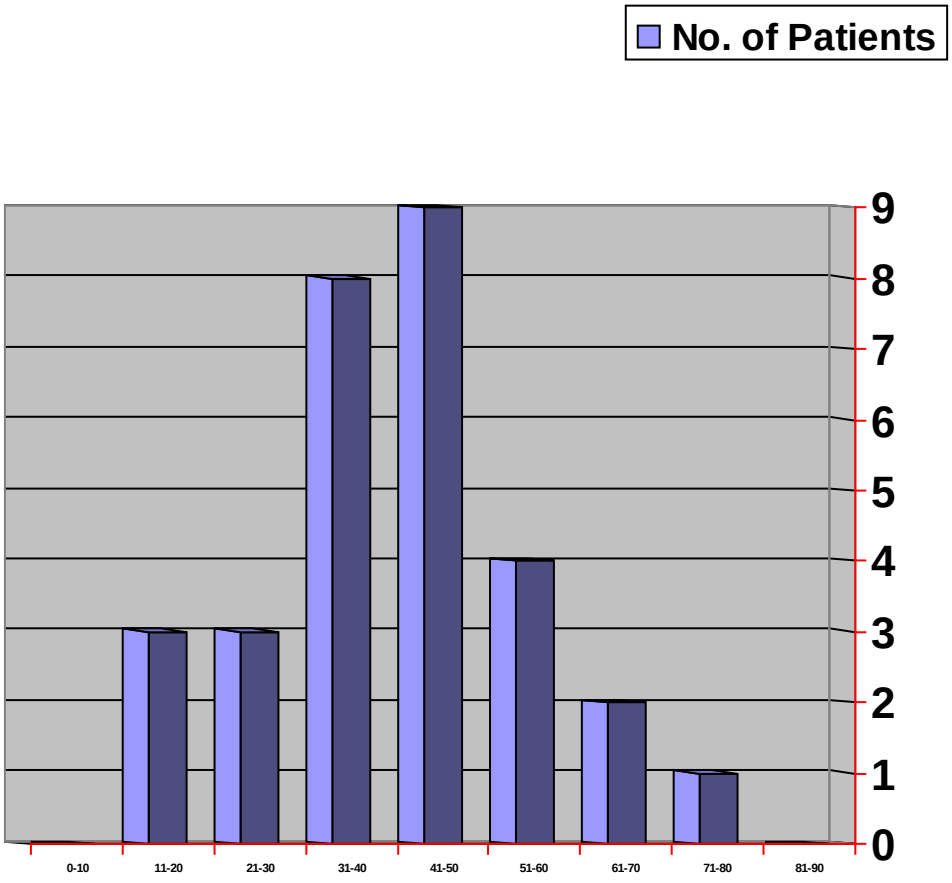
Females - 53.3%

Male: Female ratio was 8 : 7

TYPES OF LESIONS IN THE STUDY (FIG.3)

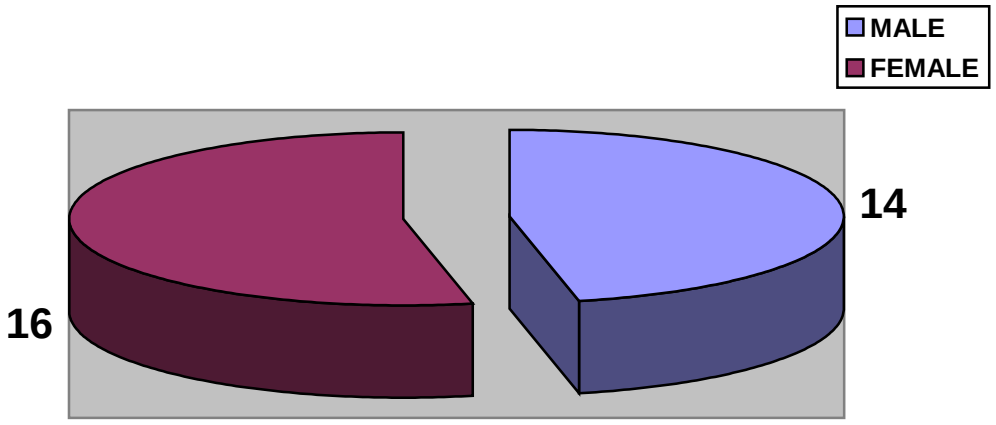
The most commonly found lesions in the study were vestibular schwannomas consisting of 24 out of 30 cases.There were four cases of meningiomas, 1 arachnoid cyst and one astrocytoma.

AGE DISTRIBUTION (Fig.1)

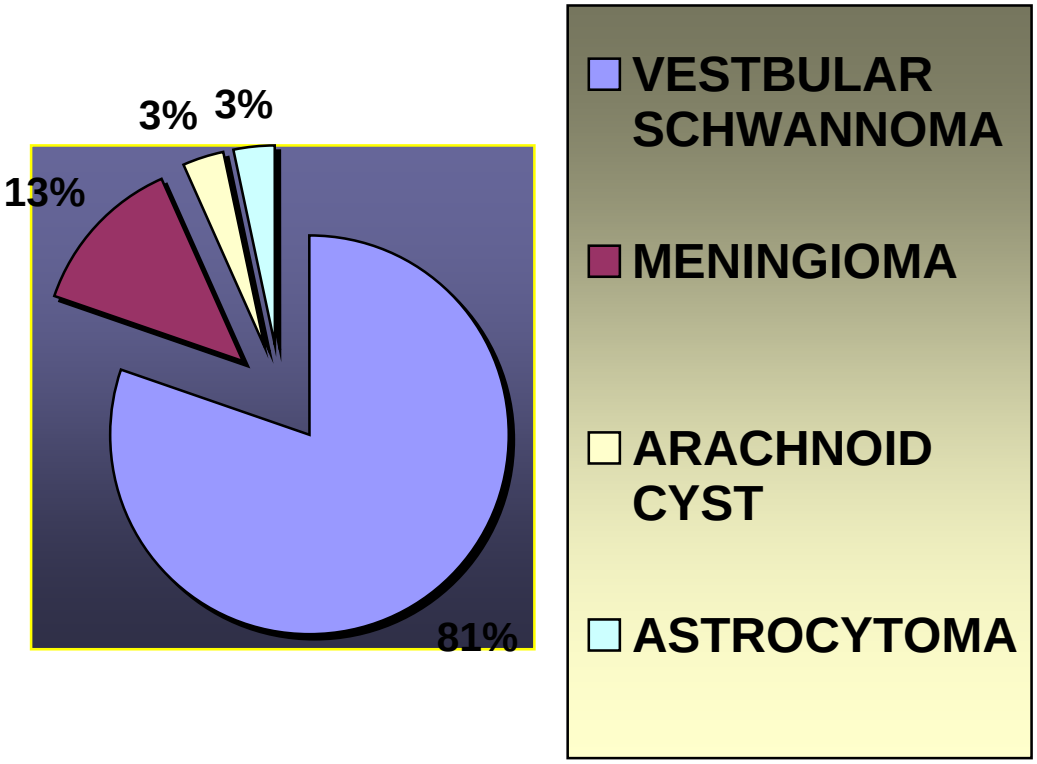


Age Group

SEX DISTRIBUTION (Fig.2)



TYPES OF LESIONS IN THE STUDY (FIG.3)



Vestibular Schwannoma	80 %
Meningioma	13 %
Arachnoid Cyst	3.33 %
Astrocytoma	3.33 %

One case had bilateral vestibular schwannoma.

SYMPTOMS (Fig.4)

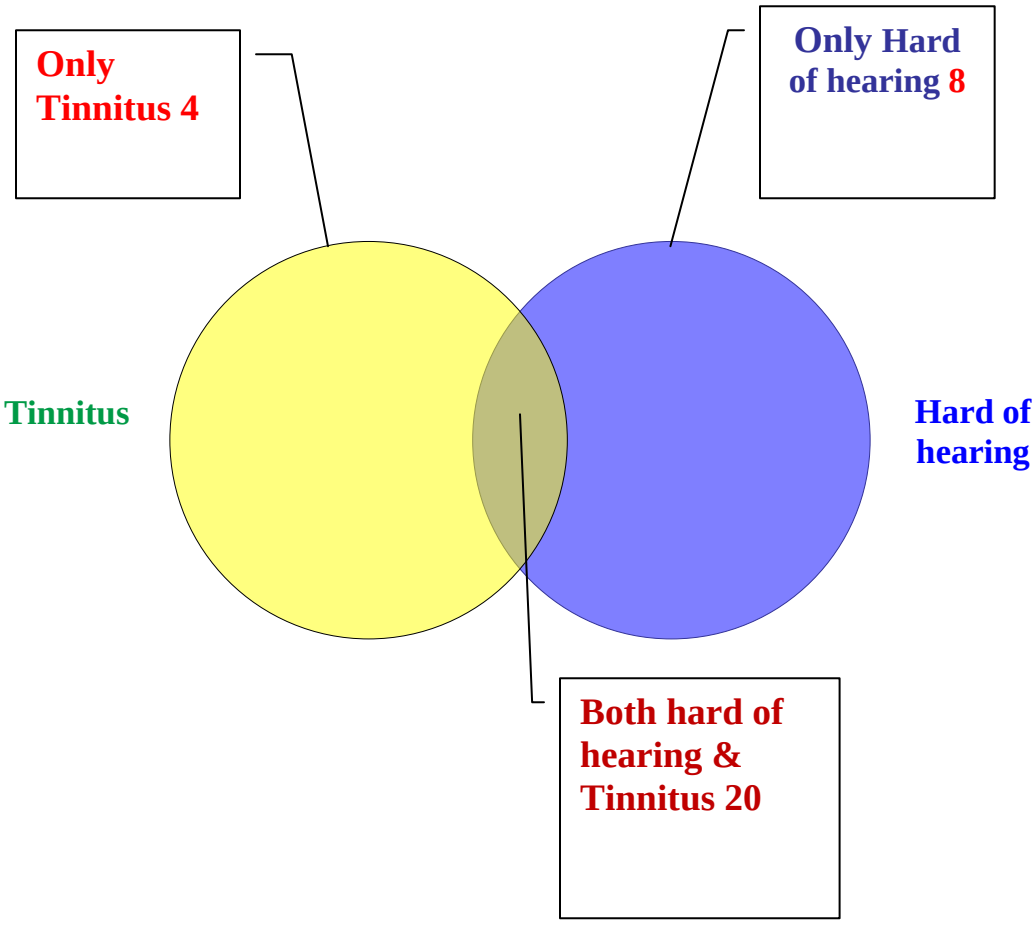
The first symptom was Hard of Hearing in 24 out of 30 cases. (80%). The first symptom was tinnitus in 6.7 % , headache in 13 %, and giddiness in 6.7% of cases.

Both Hard of Hearing and Tinnitus	66.7%
Only Hard of hearing	13%
Only Tinnitus	6.7%
Headache	40%
Hypoaesthesia of face	13.3%
Giddiness	23%
Unsteadiness	13.3%

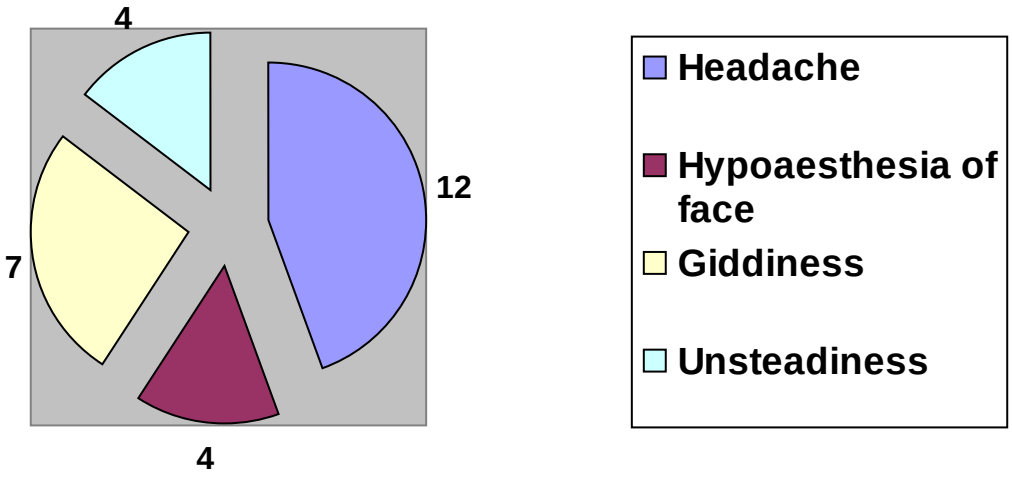
Facial Nerve was found affected in 8 cases ; 26.7% of cases.

One patient had facial tics on the side of the lesion.

SYMPTOMS: (FIG.4)



OTHERS SYMPTOMS



Two cases had symptoms of raised intracranial hypertension ; due to hydrocephalus.

THE FREQUENCY OF SYMPTOMS

The duration of symptoms ranged from 1 month to 3 years.

The mean duration of symptoms was 1.45 years.

The median duration of symptoms was 1 year and 4 months.

PURETONE AUDIOGRAM (FIG.5)

Cases with Mild Sensorineural Hearing loss were 7 (23.3%)

Cases with Moderate Sensorineural Hearing Loss were 2 (6.67%)

Cases with Severe Sensorineural Hearing Loss were 6 (20%)

Cases with Profound Sensorineural Hearing Loss were 7 (23.3%)

Cases with Total Sensorineural Hearing Loss were 7 (23.3%)

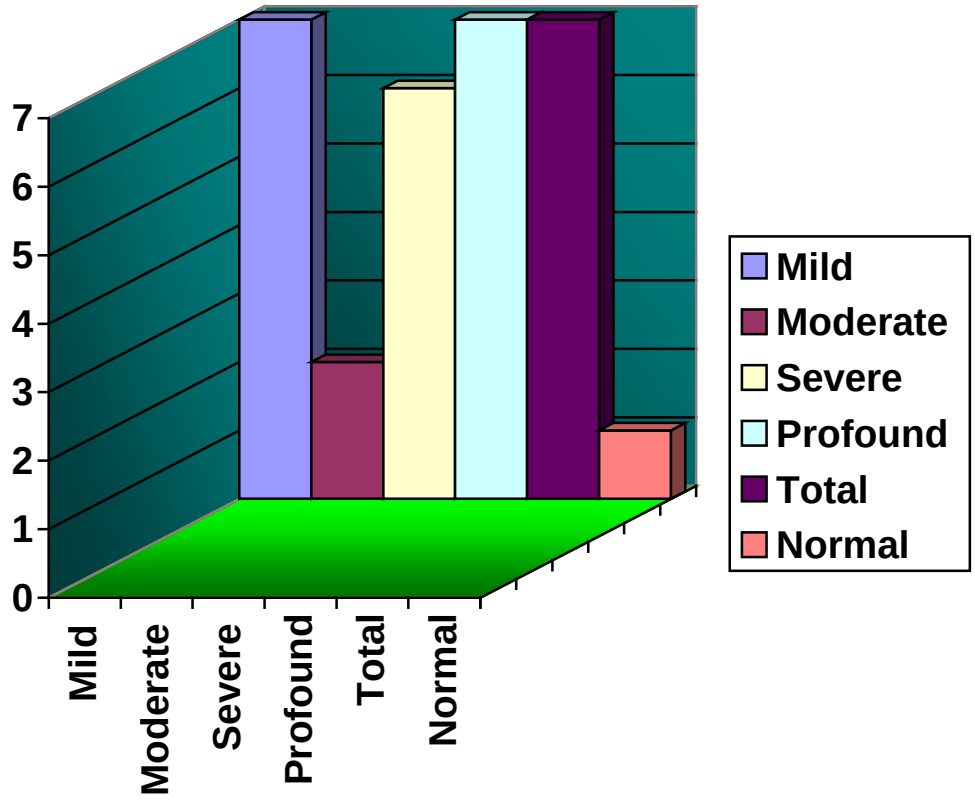
1 case (3.33%) had a normal hearing.

6 cases (20%) had a high frequency hearing loss.

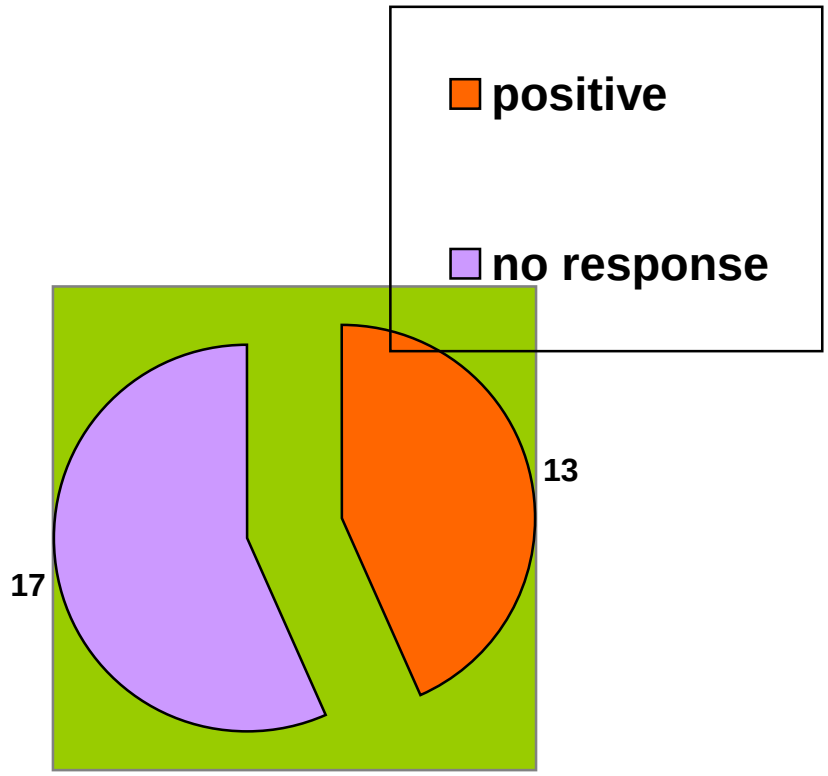
TONE DECAY TEST and SUPRATHRESHOLD ADAPTATION TEST (FIG.6)

13 cases out of 30 cases (43.3%) showed positive results ; indicating a retrocochlear pathology.

PURETONE AUDIOGRAM (FIG.5)



TONE DECAY TEST & SUPRATHRESHOLD ADAPTATION TEST (FIG.6)



These tests could not be done in 17 patients who had severe to profound hearing loss.

SHORT INCREMENT SENSITIVITY INDEX and ALTERNATE BINAURAL BALANCE TEST (FIG.7)

13 cases had a SISI score of 30 – 40%. None of the cases showed recruitement phenomenon.

In 17 cases with severe to profound hearing loss the tests could not be done.

BRAINSTEM EVOKED RESPONSE AUDIOMETRY (FIG.8)

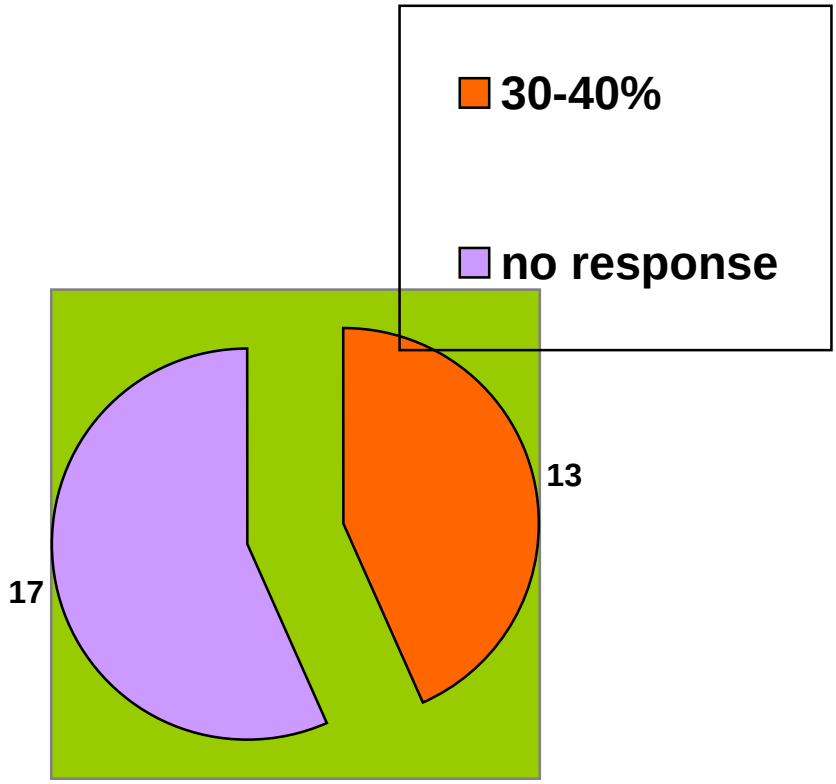
Cases with prolonged absolute latency of waves was 14 (46.67%)

Cases with prolonged interaural latency was 14 (46.67%)

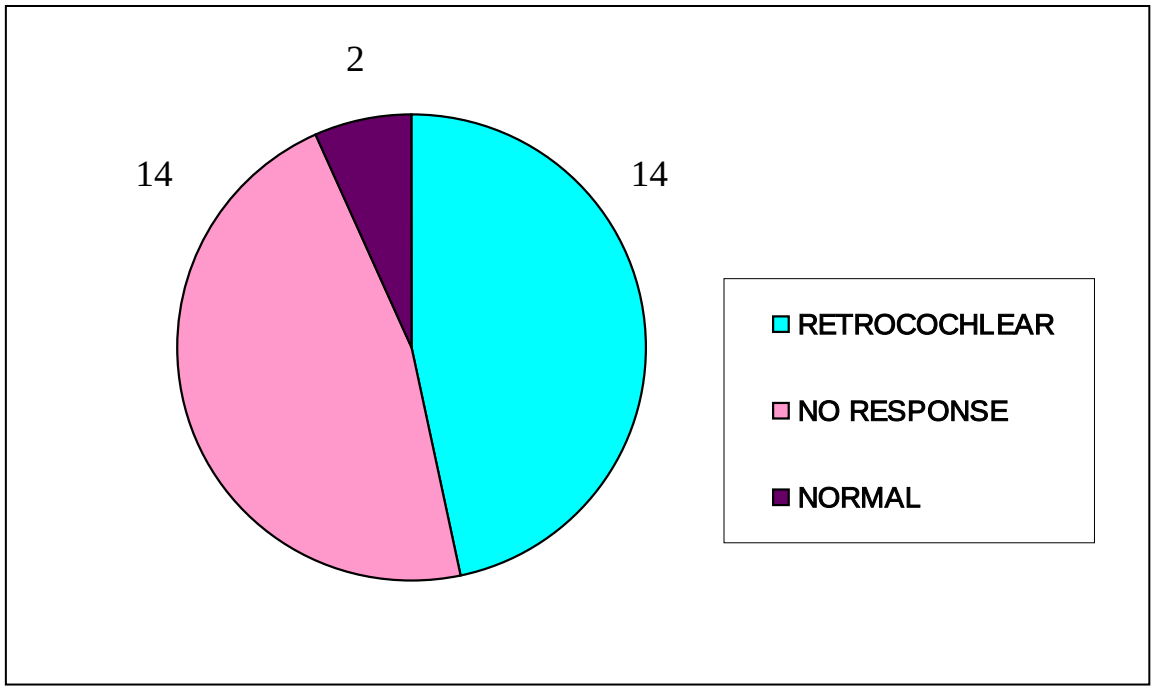
Cases with poor wave morphology were 14 (46.67%)

Due to profound hearing loss BERA showed no response in 14 cases.(46.67%)

SHORT INCREMENT SENSITIVITY INDEX (FIG.7)



BRAINSTEM EVOKED RESPONSE AUDIOMETRY (FIG.8)



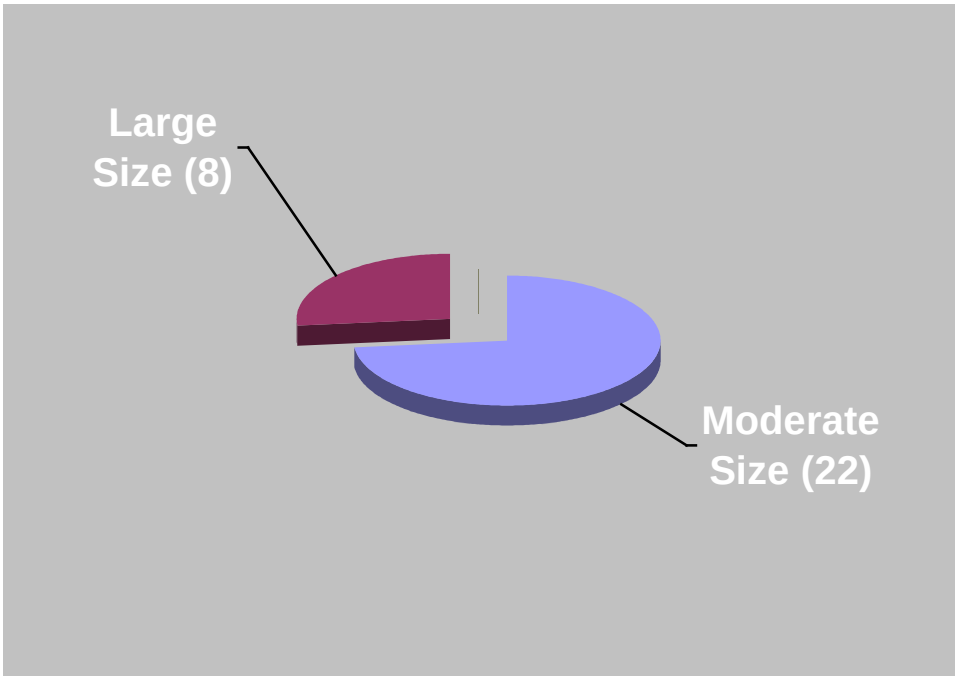
BERA showed a normal response in only two cases.(6.67%), thus showing a sensitivity of 93.3 % in detecting retrocochlear space occupying lesions

Magnetic Resonance Imaging (FIG.9)

Size of the tumours

Small tumours <1cm	Nil
Moderate size	22 Cases
Large size	8 Cases

TUMOUR SIZE: (FIG.9)



DISCUSSION

Incidence of the Tumours

Acoustic Neuromas consist of 60- 90% of all cerebellopontine angle tumours.

In this study 80% of cerebellopontine angle tumours were acoustic neuromas. Meningiomas consist of 6.8% of cerebellopontine angle tumours (valavani's et al) or 6.3% (Gonzalez Revilla)

In this study 13% of the tumours were meningiomas.

Female predilection has been noted in some studies for both acoustic neuromas and meningiomas. In this study 53.3% are females.

The average age of sporadic Acoustic Neuromas is 50 years, whereas in NF2 patient presents at a younger age with a mean age of 31 years. In this study the mean age is 43.4 years .One case in this study of bilateral vestibular schwannoma had an age of 18 years.

Regarding meningiomas the peak incidence is in the 6^{th} and 7^{th} decade of life, which in this study was found to be 58.8 years.

Symptomatology of Retrocochlear

Space Occupying Lesions

Majority of studies verify that the initial or presenting symptom is hearing loss with or without tinnitus in about 80 % of patients, which in this study is also found to be 80 %.

Tinnitus as the only presenting symptom is found in about 7 % of patients and hearing loss without tinnitus is identified in between 40% and 80%. In this study the cases with only tinnitus as the presenting symptom was found to be 6.7% and cases with hearing loss alone as the presenting symptom was 13 %.

In meningioma series tinnitus was identified in only 34% to 50% of patients at diagnosis.

The average number of years from the onset of symptoms to diagnosis is about 4 according to Selesnick & collegues in 1992[26]; which in this study was found as 1.5 years.

The time from the onset of symptoms to the diagnosis is less than 1 year in only about 20 % of patients, which was found as 26 % in this study.

In various studies , 3% -22% of patients have three frequency Puretone average threshold better than 20- 25dB and 16% - 45%of patients have Puretone average less than 70dB. In this study, 3.3% had Puretone average better than 20-25dB and 53.3 % had Puretone averages less than 70 dB. 46.6 % were found to have a Puretone average above 70 dB.

The symptoms of hearing loss or tinnitus are independent of tumour size but there is a definite trend toward normal hearing in tumour less than 1 cm.

Speech discrimination for the eighth nerve tumour patients is quite poor. Selesnick and coworkers[26] found that patients with less than 1cm in diameter had an average puretone loss of 40dB and 50% of those patients had a SDS of greater than 81% and the series of less than 2cm tumour of Dornhoffer[27] found 37% with normal speech discrimination. In patients with medium and large tumours , the average speech reception threshold was 47 and 58dB respectively and only 25% of patients with medium a large tumours had an SDS better than 81%.

In this study, speech audiometry and special tests could be done only in 43% of cases, the rest having severe-total hearing loss, and a result indicating a retrocochlear pathology could be obtained in all the 43% cases.

The majority of series demonstrate about a 5% to 7% incidence of truly normal hearing , which in this study was 3.33%

Brainstem Evoked Response Audiometry

Selters and Brackmann, House and Brackmann and Clemis and McGee in 1970s demonstrated 92% - 98% sensitivity for absent or abnormal Auditory Brainstem Response in Acoustic Neuroma patients. Auditory Brainstem Response shows a consistent asynchronous retrocochlear pattern in tumours more than 2 cm but in intracanalicular tumours, a much greater percentage will show a normal pattern.

Grabel et al in 1991[28] stated that 30% of intracanalicular tumours showed a normal Auditory Brainstem Response.

Gordon in 1995[29] stated that 31% of intracanalicular tumours and 15.5% of tumours less than 1.5cm showed a normal Auditory Brainstem Response.

Brainstem Evoked Response Audiometry in other retrocochlear lesions:

Laird and coworkers[30] and Granick and collegues[31] had a series of meningiomas of the posterior fossa of all of which showed a positive Auditory Brainstem Response.

House and Brackmann[32] demonstrated that only 75% of cerebellopontine angle lesions that were not acoustic neuromas had an abnormal Acoustic Brainstem Response.

Brainstem evoked response audiometry showed results corresponding to retrocochlear pathology in 46.7% of cases and a normal result in 6.67% of cases. Due to hearing loss more than 70 dB, Brainstem evoked response audiometry showed no response in the remaining 46.7%, hence being sensitive upto only 93% ; two cases which showed normal Brainstem evoked response audiometry waves were posterior cranial fossa meningiomas.

Marangos and coworkers[33] found 23.5% of meningiomas had a normal Auditory Brainstem response.

MAGNETIC RESONANCE IMAGING

The sensitivity of contrast enhanced T_1 – weighted MR imaging can approach 100% in detecting schwannomas[31] .

In the study, all the cases were diagnosed by contrast enhanced MRI.

CONCLUSION

A high index of suspicion needs to be maintained when symptoms of unilateral hearing loss and tinnitus co-exist in a patient. The patient then has to undergo a battery of audiological tests to rule a retrocochlear lesion.

The first line audiological investigation is the PureTone Audiometry, but which cannot differentiate a cochlear from a retrocochlear lesion.

The speech audiometry and special tests (SISI, TDT, ABLB etc.) are useful in differentiating cochlear from retrocochlear lesions. They have limitations when it comes to patients with hearing loss more than 60 dB.

The sensitivity of Brainstem Evoked Response Audiometry in detecting retrocochlear lesion is found to be 93% in this study, with an advantage of detecting the site of the lesion. This test also has limitations when it comes to patients with hearing loss more than 60dB.

No audiological test is 100 % sensitive in detecting retrocochlear lesions but when applied together the chance of early detection is increased.

The sensitivity of Gadolinium Enhanced Magnetic Resonance Imaging reaches almost 100%, thus leaving it as the gold standard investigation for diagnosis of the condition.

Taking into account of the cost factor of Gadolinium enhanced Magnetic Resonance Imaging, it cannot be used as a screening test, whereas Brainstem Evoked Response Audiometry can be used to screen for retrocochlear space occupying lesions when there is a high index of suspicion.

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PROFORMA FOR OTONEUROLOGICAL DIAGNOSIS OF RETROCOCHLEAR LESIONS

S.No:
Name:
Address:
Hospital No:
Sex:
Date of study:
Age:
Total Duration of Symptoms:
First Symptom:
Symptoms: Duration:
Vestibulocochlear
Tinnitus
- Ipsilateral/Contralateral
- Continuos/Episodic

Hearing Loss
- Unilateral/Bilateral
- Sudden/Gradual

Vertigo
- Episodic/Continuous
- Vomiting
- Frequency of attacks
- Duration

Facial Nerve:
- Weakness
- Taste

Trigeminal Nerve:
- Sensory impairement
- Motor symptoms

Cerebellar:
- Unsteadiness
- Incordination
- Dysarthria

Others
- Headache
- Headache with vomiting
- Drowsiness
- Nasal regurgitation

Change of voice
Neurological Examination:
7th nerve
House and Brackmann Grading
Taste
8th Nerve
Rinne
Weber
5th Nerve
Sensory: V1
V2
V3
Motor:
Corneal Reflex
9th and 10th
Cerebellar:
Nystagmus
Truncal Ataxia
Appendicular Ataxia
Pyramidal Signs
Others:
Audiological Tests:
Pure tone audiometry: High Frequency Loss
Low Frequency Loss
BERA:
Normal
Retrocochlear
No Response
MRI

MASTER CHART

S. No.	Hosp.No	Name	Age	Sx	Dur	S1	HL	TI	GD	FS	GT	OS	PTA	SDS	TDT, STAT	SISI	BERA	MRI
1	658/08	Neelavathy	58	F	7m	HL	Y	Y	Y	N	N	FP	SNHLSev	<30%	+ve	30%	NR	AN
2	836/08	Sumathi	26	M	1m	HL	Y	Y	N	N	N	HF	SNHLMild	<30%	+ve	30%	RC	AN
3	1029/08	Ravichandran	50	M	6m	HL	Y	Y	Y	N	N	HA	SNHLProf	-	_	_	RC	AN
4	2660/08	Johnson	20	M	1.5Yr	HL	Y	Y	N	N	N	HA	SNHLSev	<30%	+	30%	NR	AN
5	3661/08	Shanthi	40	F	2Yr	HL	Y	Y	N	Y	N	_	SNHLMod	<30%	+ve	30%	RC	AN
6	5695/08	Rajendran	50	M	2Yr	TI	Y	Y	N	N	N	_	SNHLMild	<30%	+ve	30%	RC	AN
7	7605/08	Vijaya	30	F	1.5Yr	HL	Y	Y	N	N	N	_	SNHLSev	-	_	_	RC	AN
8	34214/08	Lakshmi	50	F	3Yrs	GD	Y	Y	Y	N	N	HA	HFSNmild	<30%	+ve	30%	RC	AC
9	36123/08	Murugyan	35	M	1Yr	TI	Y	Y	Y	N	N	_	SNHLProf	-	_	_	NR	AN
10	40544/08	Amuda	46	F	3Yrs	HA	N	N	Y	Y	N	HA	HFSNmild	<30%	+ve	20%,	RC	AN
11	46347/08	Umapathy	31	M	2Yrs	HL	Y	Y	Y	N	N	_	SNHLProf	-	_	_	RC	AN
12	47222/08	Thilaka	32	F	2Yrs	HL	Y	Y	Y	N	N	US	HFHLmild	<30%	+ve	30%	RC	AN
13	48564/08	Pachiappan	67	M	6m	HL	Y	Y	Y	N	N	_	Nl HG	<30%	+ve	30%	RC	MG
14	53218/08	Vijaya	46	F	1.5Yrs	HL	Y	Y	N	N	N	HA	SNHLsev	-	_	_	RC	AN
15	62814/08	Rajendran	18	M	1Yr	HL	Y	Y	N	N	N	_	SNHLProf	-	_	_	NR	AN-BL

S.No.	Hosp.No	Name	Age	Sex	Dur	S1	HL	TI	GD	FS	GT	OS	PTA	SDS	TDT, STAT	SISI	BERA	MRI
16	83665/08	Govindaraj	67	M	6m	HA	N	N	N	N	N	_	HFSNmild	<30%	+ve	30%	NL	MG
17	86933/08	Rajeswari	50	F	1.5Yr	HA	Y	N	N	Y	N	_	SNHLTot	-	_	_	RC	AN
18	618/09	Vasuki	39	F	3Yr	HL	Y	Y	Y	N	N	_	SNHLTot	-	_	_	NR	AS
19	871/09	Venkatesan	44	M	1Yr	HL	Y	Y	Y	N	N	_	SNHLSev	<30%	+ve	30%	NR	AN
20	969/09	Arunkumar	32	M	2Yr	HL	Y	Y	N	N	N	_	SNHLTot	-	_	_	NR	AN
21	1597/09	Muraleedaran	45	M	6m	HL	Y	Y	Y	Y	N	US	SNHLSev	<30%	+ve	30%	RC	AN
22	1627/09	Geetha	29	F	2Yr	HL	Y	Y	N	N	Y	_	SNHLProf	-	_	_	NR	AN
23	2642/09	Violet	58	F	2.5Yr	HL	Y	Y	N	N	N	HA	SNHLMod	<30%	+ve	30%	NL	MG
24	3678/09	Kousalya	19	F	2Yr	HL	Y	Y	N	N	N	_	SNHLTotal	-	_	_	NR	AN
25	4435/09	Balasubramaniam	55	M	1Yr	HL	Y	Y	N	N	N	_	SNHLTot	-	_	_	NR	AN
26	4675/09	Dhanabakiyam	72	F	6m	TI	Y	Y	N	N	N	_	SNHLTotal	-	_	_	NR	AN
27	7044/09	Thamimansari	40	M	1Yr	HL	Y	Y	N	N	N	HA	SNHLProf	-	_	_	NR	AN
28	8614/09	Kousalyam	39	M	1Yr	HL	Y	Y	Y	N	N	N	SNHLProf	-	_	_	NR	AN
29	46161/09	Karimullah	50	M	3Yr	HA	Y	N	N	N	N	HA	SNHLTot	-	_	_	NR	AN
30	86933/09	Rajeswari	55	F	1.5Yr	HL	Y	Y	N	N	N	_	SNHLTot	-	_	_	NR	AN

LEGEND FOR MASTER CHART

S1:First Symptom
HL:Hearing Loss
TI:Tinnitus
GD:Giddiness
FS:Facial Sensory Abnormality
GT:Gait Abnormality
OS:Other Symptoms
HA:Headache
FP:Facial Palsy
HF:Hemifacial Spasm
US:Unsteadiness
PTA:Puretone Audiogram
SNHL:Sensorineural Hearing Loss
Mild
Mod- Moderate
Sev-Severe
Prof-Profound
SDS-Speech Descrimination Score
TDT:Tone decay Test
STAT:Suprathreshold Adaptation Test
SISI:Short Increment Sensitivity Index
BERA:Brainstem Evoked Response Audiometry
NR:No Response
RC:Retrocochlear
NL:Normal
AN:Acoustic Neuroma
MG:Meningioma
AC:Arachnoid Cyst
AS:Astrocytoma

PATIENT CONSENT FORM

Study Detail: Comprehensive Study of Audiological Evaluation of Retrocochlear Space Occupying Lesions

Study Centre : Upgraded Institute of Otorhinolaryngology,
Madras Medical College & Government General Hospital, Chennai -3
Institute of Child health, Egmore, Chennai 600 008.

Patient Name :
Patient Age :
Identification Number :

Patient may tic (✓) these boxes

I confirm that I have understood the purpose of procedure for the above Study. I have the opportunity to ask the question and all my questions and doubts have been answered to my satisfaction. ☐

I understand that my participation in the study is voluntary and that I am free to withdraw at anytime without giving any reason, without my legal rights being effected. ☐

I understand that Investigator, Regulatory authorities and the Ethics committee will not need my permission to look at my health records both in respect to the current study and any further research that may be conducted in relation to it, even if withdraw fro the study. I understand that my identity will not be revealed in any information released to third parties or published, unless as required under the law. I agree not to restrict the use of any data or results that arise from the study. ☐

I agree to take part in the above study and to comply with the instructions given during the study and faithfully co operative with the study team and to immediately inform the study staff if I suffer from any deterioration in my health or well being or any unexpected or unusual symptoms, ☐

I hereby con sent to participate in this study. ☐

I hereby give permission to undergo complete clinical examination and diagnostic tests including hematological, biochemical, radiological tests. ☐

Signature/Thumb Impression: Place Date
Patient Name and Address:

Signature of the Investigator: Place Date
Study Investigator's Name:

INSTITUTIONAL ETHICAL COMMITTEE
MADRAS MEDICAL COLLEGE,CHENNAI-600 003.

Telephone : 25363970

Fax :044 – 253-5115
:044 25363970

L.Dis.No. 14597/ME5/EthicsDean/MMC/2009 Dated .09.2009

Title of the work : A comprehensive study on audiological evaluation of retrocochlear space occupying lesion
Principal Investigator :
Department : Dr. Nitha Neelambaran; PG-M.S(ENT) madras medical college - ch-3.

The request for an approval from the Institutional Ethical Committee(IEC) was considered on the IEC meeting held on 23rd September 2009 at 2.00P.M. in Madras Medical College, Deans, Chamber, Chennai-3. /pharmacology seminar hall. madras medical college -ch-3.

The members of the Committee, the Secretary and the Chairman are pleased to approve the proposed work mentioned above, submitted by the principal investigator.

The principal investigator and their term are directed to adhere the guidelines given below:

1. You should get detailed informed consent from the patients/participants and maintain confidentiality.
2. You should carry out the work without detrimental to regular activities as well as without extra expenditure to the Institution or Government.
3. You should inform the IEC in case of any change of study procedure, site and investigation or guide.
4. You should not deviate form the area of the work for which I applied for ethical clearnance.
5. You should inform the IEC immediately, in case of any adverse events or serious adverse reactions.
6. You should abide to the rules and regulations of the institution(s).
7. You should complete the work within the specific period and if any extension of time is required, you should apply for permission again and do the work.
8. You should submit the summary of the work to the ethical committee on completion of the work.
9. You should not claim funds from the Institution while doing the work or on completion.
10. You should understand that the members of IEC have the right to monitor the work with prior intimation.

SECRETARY
IEC, MMC,CHENNAI

CHAIRMAN
IEC MMC CHENNAI

21/10/09

DEAN
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