Ramamurthi & Tandon's

MANUAL OF NEUROSURGERY

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PREFACE

In the era of rapid advances in knowledge, techniques and technologies, it is difficult for a textbook to remain contemporary for any length of time. On the other hand, timely updating requires herculean effort, especially as it entails cooperation of a large number of contributors of a multi-authored book.

Keeping in view the above, we have attempted a new experiment of bringing out this concise edition of the book in a totally new format, i.e. converting the text in bulleted form, retaining the essential information but excluding the supportive evidence provided in the detailed textbook. At the same time, opportunity is taken to add recent advances. The publication of our *Textbook of Neurosurgery (Third Edition)* and the *Textbook of Operative Neurosurgery (First Edition)* has resulted in an abridged version of the voluminous material.

The book is divided into XVII sections covering all important areas of neurosurgery, e.g. diagnostics, congenital malformations, head and spinal injuries, infections, vascular malformations, central nervous system tumours, etc. In addition, there are chapters on special neurosurgical techniques, such as stereotactic and functional neurosurgery, surgery for epilepsy, movement disorders, psychosurgery and pain. Each section has a number of sub-sections, for example, the section on infections includes brain abscess, subdural empyema, tuberculosis, fungal infections, cysticercosis, etc. Each sub-section provides stand-alone comprehensive information on the subject. This format of the text, as well as the multiple authorship of the book, has no doubt resulted in some duplication of information. Nevertheless, enormous amount of information has been restricted to a manageable number of pages.

We hope, the students and ever busy practicing neurosurgeons will find this new format as an aide-memoir for ready reference. For greater details, reference can be made from the full-text editions of our earlier textbook.

We take this opportunity to profusely thank all the authors, who notwithstanding their extremely busy life provided their valuable, updated contributions in the new format. Special thanks are due to Shri Jitendar P Vij (Group Chairman), Mr Ankit Vij (Managing Director) and Tarun Duneja (Director-Publishing) of M/s Jaypee Brothers Medical Publishers (P) Ltd, New Delhi, India, for their cooperation and the excellent quality of this publication.

Prakash Narain Tandon
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CHAPTER 1

Electrodiagnosis

Manjari Tripathi • Priya Gupta • Sarat Chandra P

NERVE CONDUCTION STUDIES

- Physiologically, the nerve transmits an electrical impulse across its axon both antidromically and orthodromically.
- Each stimulus has a proximal as well as distal transmission. This property is used to assess compound muscle action potential (CMAP), sensory nerve action potential (SNAP), F-response, H-reflex and SSR.
- Each potential is described by its latency, amplitude, area and configuration.
- To obtain the velocity, the nerve is stimulated at two different sites and recording is done on the same muscle, and the difference in response latency is multiplied by the distance between these two stimulation sites.

Definition of Related Terms

- ***Latency:*** This is the time in milliseconds from the application of a stimulus to the initial deflection from the baseline, either positive or negative, and is the time required for the action potentials in the fastest conducting fibres to activate the muscle fibres.
- ***Conduction velocity:*** It is calculated by measuring the difference in distance and latency between sites of stimulation in metres per second.
- ***Amplitude:*** It is measured in height in millivolts from the baseline to the peak of the negative phase (upward deflection). It is proportional to the number of muscle fibres activated and gives an estimate of the amount of functioning nerve and muscle.
- ***F-response:*** It results from the backfiring of antidromically activated anterior horn cells. It is especially helpful in assessment of motor conduction of the proximal segment of a stimulated axon along with the entire length of the peripheral axon.
- ***H-reflex:*** It is an electrically elicited spinal monosynaptic reflex and bypasses the muscle spindles. The group 1A sensory fibres and alpha motor neurons form the respective afferent and efferent arcs of this reflex. The H-reflex latency and amplitude are helpful in the diagnosis of mild-S1 radiculopathy.

Types of Neuropathy

The following are the types of neuropathy:

- Demyelinating neuropathy
- Axonal neuropathy
- Mixed demyelinative/Axonal neuropathy.

Important Points to Remember

- Do not trust electrophysiological studies (EPS) if they do not match your clinical examination finding.
- The role of EPS is to corroborate clinical history and examination, and is not a substitute for the latter.
- While interpreting nerve conduction study (NCS), normative data of the concerned laboratory should be available before interpretation.

ELECTROMYOGRAPHY

- The pattern of electrical activity in muscle [i.e. electromyography (EMG)], both at rest and during activity, may be recorded from a needle electrode inserted into the muscle.
- The nature and pattern of abnormalities relate to disorders at different levels of the motor unit.
- Before planning to start EMG, the clinician should have relevant NCS and clinical details of the patient.

Electromyography Findings

At rest

- Insertion activity
- Spontaneous activity.

At voluntary movements

- Assessment of individually recruited motor unit action potential (MUAP)
- Size
- Shape
- Stability
- Assessment of activation pattern of MUAPs
- Recruitment
- Interference pattern (IP).

Insertional Activity

It lasts for a few hundred milliseconds, usually 300–500 ms. Appear as a cluster of positive or negative repetitive high-frequency spikes.

Increased insertional activity indicates instability of the muscle membrane indicating:

- Denervation, usually an early finding 1–2 weeks after nerve injury
- Myotonic disorders
- Necrotising myopathies, such as inflammatory myopathies.

Decreased insertional activity suggests either fibrotic or severely atrophied muscles or functionally unexcitable muscles (e.g. during attacks of familial periodic paralysis).

Spontaneous Activity

Normal

- Normally no spontaneous activity is seen except at the motor endplate region.
- Two types of normal endplate spontaneous activity are present.
- ***Endplate noise:*** These are extracellularly recorded miniature endplate potentials and non-propagating depolarisations. It is caused by spontaneous

release of acetylcholine quanta. These are negative potentials, irregular in rhythm, with amplitude of 10–50 uV and duration of 1–2 milliseconds (sounds like a "sea shell held to the ear").

- ***Endplate spikes:*** These are discharges of single muscle fibres generated by activation of intramuscular nerve terminals irritated by the needle. They may originate in the intrafusal muscle fibres. They are irregular having an initial negative deflection, firing at 5–50 impulses per second, with amplitude of 100–200 pV with duration of 3–4 milliseconds (sounds like "clacking or buzzing").

Abnormal

- Fibrillation potentials
- Fasciculation potentials
- Complex repetitive discharges
- Myotonic discharges
- Myokemic discharges
- Neuromyotonic discharges
- Cramps.

Fibrillation potentials

- These are spontaneous action potentials of denervated single muscle fibres.
- They result from the resting membrane potential of the denervated fibre and usually appear 10–21 days after muscle denervation.
- They are brief spikes (usually triphasic with initial positivity) or positive waves (initial positivity with slow negativity, sawtooth appearance), fire regularly at 1–30 Hz, with an amplitude of 20–200 pV; gradually decreasing with time, with a duration of 1–5 milliseconds (sounds like "crisp and clicking" and "tick-tock of a clock").

Causes

- Denervation, useful in localising lesions affecting motor neurons, spinal roots, plexus or peripheral nerve.
- May persist in paraspinous muscles for years after surgery.
- Inflammatory myopathy.

Grading system is used (from 0 to 4) to semiquantitate fibrillation potentials

0	–	no fibrillations
+1	–	persistent single trains of potentials (> 2 seconds) in at least two areas
+2	–	moderate number of potentials in three or more areas
+3	–	many potentials in all areas
+4	–	abundant spontaneous potentials nearly filling the oscilloscope.

Fasciculation potentials

- Fasciculation potentials are spontaneous discharges of a motor unit.
- They originate from the motor axon anywhere along its length.
- Their size and shape are like MUAPs, have an irregular rhythm and a lower firing rate than voluntary MUAPs (sounds like "corn popping").

Causes

- Diseases of anterior horn cells (MND)
- Radiculopathies
- Entrapment neuropathies
- Peripheral polyneuropathies

- Cramp fasciculation syndrome
- *Others*: tetany, thyrotoxicosis and overdose of anticholinesterase medication
- They may also occur in healthy people.

Complex repetitive discharges

- These result from the nearly synchronous firing of a group of muscle fibres.
- One fibre in the complex serves as a pacemaker, driving one or several other fibres so that the individual spikes in the complex fire in the same order as the discharge recurs.
- One of the late-activated fibres re-excites the principal pacemaker to repeat the cycle. They are shaped containing 10 or more distinct spikes, regular in rhythm, firing at 5–100 Hz, typically begin abruptly, with an amplitude of 50 pV–1 mV, with a duration of 50–1000 milliseconds (sounds like "machine gun" on the loudspeaker).
- Complex repetitive discharges are non-specific and seen in many chronic disorders, including chronic neuropathies and myopathies.
- They may also be found in the iliopsoas or cervical paraspinal muscles of apparently healthy people, probably implying a clinically silent neuropathic process.

Myotonic discharges

- Abnormal insertional activity of recurring single-fibre potentials wax and wane in the range of 10 LIV–1 mV, varying inversely with the firing rate of 20–150 Hz (accelerating or decelerating motorcycle or chainsaw).
- Myotonic discharges may occur with or without clinical myotonia in the myotonic dystrophies (types I and II) and other myopathies.

Myokymic discharges

- These originate ectopically in motor nerve fibres and result from complex bursts of grouped repetitive discharges in which motor units fire repetitively.
- Usually with 2–10 spikes with a firing rate of 30–40 Hz, each burst recurs at regular intervals of 1–5 seconds (sounds like "marching soldiers" on the speaker).

Neuromyotonic discharges

- Here, the muscle fibres fire repetitively at high frequency (150–250 Hz) and produce a pinging sound on the speaker.
- They continue during sleep and diminish in intensity with distal nerve blocks.

Cramp discharges

- These include sustained involuntary muscle contraction during cramps.
- The discharge consists of MUAPs, with a firing rate of 40–60 Hz, with abrupt onset and cessation.

MOTOR UNIT ACTION POTENTIALS

- The MUAP is the extracellular electrode recording of a small portion of a motor unit.
- They are triphasic in shape, measured as total peak-to-peak amplitude.
- It normally varies from several hundred microvolts to a few millivolts.
- ***Duration:*** It normally varies from 5–15 milliseconds. Long-duration MUAPs with high amplitude are the best indicators of reinnervation. Short-duration MUAPs with low amplitude are seen in disorders associated with loss of muscle fibres, like myopathies.

- ***Phase:*** This is the deviation of the signal from its beginning until its return to the baseline. An MUAP having more than four phases is considered polyphasic. Increased polyphasics is a nonspecific abnormality seen both in myopathies and neuropathies.
- ***Late component (satellite or linked potential):*** This is a time locked waveform to the main MUAP, but separated from it by an isoelectric interval. It implicates early reinnervation of muscle fibres by collateral sprouts from adjacent motor units.
- ***Complexity:*** The MUAP with greater than 4 phases or a satellite potential is said to be complex.
- ***Variability:*** These include changes in shape of the MUAP on consecutive discharges. It indicates deficient neuromuscular transmission. They may indicate NMJ disorder, MND, polyneuropathy or radiculopathy and early stage of reinnervation.

Electrodiagnostic Findings in Nerve Injury

The electrodiagnostic findings (EDX) provide information about:
- Site of the injury
- Underlying pathophysiology
- Rate of recovery.

Neuropraxia

- No axonal degeneration, only conduction block: focal site of demyelination.
- Aetiology: Compression or traction injury.
- Large myelinated fibres are more susceptible to compression and ischaemia (motor).

Electrodiagnostic findings
- Nerve conduction is normal distally, but altered across the injury site (consider axonal loss if amplitude decreased proximally and distally).
- Needle EMG shows decreased recruitment, but no abnormal spontaneous potentials.
- Normal conduction returns in days/weeks (due to remyelination of the damaged segment).

Axonotmesis

- Axonotmesis implies axon damage with preservation of the endoneurium, perineurium and epineurium, and there will be Wallerian degeneration of the axon.
- Motor NCS lost day 4–7 (NMJ fragmentation).
- Sensory NCS lost day 8–10.
- Preservation of endoneurium allows for regeneration with reinnervation.
- Recovery time dependent on distance for reinnervation.

Electrodiagnostic findings
- Day 0–3: same as neuropraxia.
- Day 4–7: decreased motor amplitude.
- Day 8–10: decreased sensory amplitude.
- Day 10–14: abnormal spontaneous potentials on EMG (PSW, Fibs).
- Month 6–12: “nascent pot’s (S > M) and “jitter”.
- Performing EDX too early may lead to misleading information (wait 2–4 weeks).

- An early sign of axonotmesis is decreased CMAP amplitude (30–40% lower than the contralateral side).
- Repeat testing will be required in about 2–3 weeks.

Neurotmesis

- Disruption of axon, endoneurium and connective tissue (perineurium and epineurium).
- Poor prognosis for reinnervation.
- EDX finding are the same as above.

SENSORY EVOKED POTENTIALS

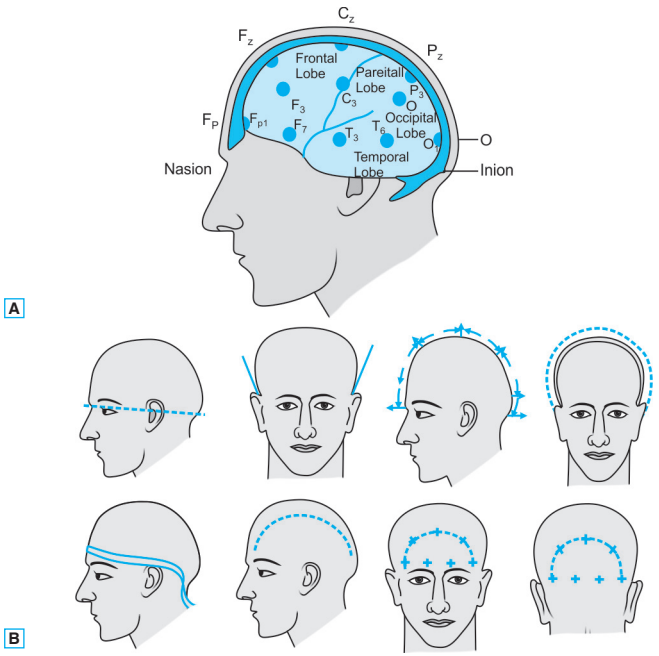
- Recording of spinal or cerebral potentials elicited by stimulation of specific afferent pathways helps in monitoring the functional integrity of these pathways, although it does not indicate the pathologic basis of lesions involving them.

Somatosensory-evoked Potentials

- The somatosensory-evoked potentials (SEPs) are obtained by multiple electrical stimulations of peripheral nerves.
- The electrical signals generated are measured at the level of the peripheral nerves (over the brachial plexus or popliteal fossa), spinal cord (lumbar as well as high cervical), and cortex (scalp).
- These recordings are made over the scalp and skin on the neck and the back (over the spinous processes of the vertebrae) to monitor these signals in the cortex and the spinal cord.
- This method relies mostly on the stimulation of the large myelinated somatosensory fibres, which transmit the impulses to the spinal cord by the posterior column system.
- Median nerve stimulation at the wrist or posterior tibial nerve stimulation at the popliteal fossa or medial malleolus (ankle) is commonly used.
- Electrode sites on the scalp are marked using the 10–20 international system (Figs 1A and B). The recording sites preferred by the authors are C3, C4, CZ, FPZ, FZ, A1 and A2.
- The recommended stimulus is a monophasic pulse of 10–20 mA and 100 μs duration.
- The stimulus is applied to the median nerve at the wrist or the posterior tibial nerve at the ankle near the site where the nerve passes posterior to the medial malleolus. Spinal cord intra-operative SEP monitoring often involves the posterior tibial nerve; but, when surgery is above the eighth cervical spinal cord level, then median nerve intraoperative SEP monitoring is used.

Role of Somatosensory-evoked Potential in Neurosurgical Diseases

- Disorders of the peripheral nerves
- Plexus lesion
- Radiculopathy
- Thoracic outlet syndrome
- Cervical myeloradiculopathy
- Malingering
- Intraoperative assessment of the functional integrity of neural pathways.



Figs 1A and B: (A) Actual representative areas of the brain in the 10–20 system. (B) Landmarks and measurements of the 10–20 system

Disorders Associated with Unrecordable Intraoperative SEPs Include

- The most frequent conditions leading to the problem of poor data acquisition are neural tube defects and severe spastic quadriparesis with atrophy of the lower extremities.
- Sequelae of spinal cord trauma
- Advanced spondylosis with myelopathy
- Scoliosis with myelopathy
- Peripheral neuropathy
- Spinal cord tumour.

BRAINSTEM AUDITORY- EVOKED POTENTIALS

Brainstem auditory-evoked potentials (BAEPs) are elicited by monaural stimulation with repetitive clicks and are recorded between the vertex of the scalp and the mastoid process or earlobe (Fig. 2).

The presence, latency and interpeak latency of the first five positive potentials recorded at the vertex are evaluated.

Brainstem Auditory-evoked Potential Wave Generators

Wave I : Distal acoustic nerve
Wave II : Proximal acoustic nerve/cochlear nucleus
Wave III : Superior olivary complex at the level of the lower pons

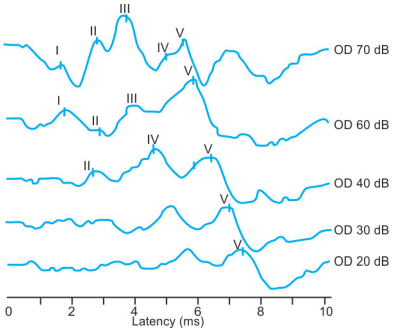


Fig. 2: Normal brainstem auditory-evoked potential wave

Wave IV : Lateral lemniscus
Wave V : Inferior colliculus or upper pons

- The findings are helpful in screening for acoustic neuromas, detecting brainstem pathology and evaluating comatose patients.
- The BAEPs are normal in coma due to metabolic/toxic disorders or bihemispheric disease, but abnormal in the presence of brainstem pathology.
- Intraoperative BAEP monitoring has been shown to be useful for preservation of hearing and vestibular nerve function during the resection of acoustic neuroma and other posterior fossa surgeries.
- The types of surgeries in which BAEPs are also used include posterior fossa and petroclival skull-base tumours, arteriovenous malformations, aneurysms and decompressive procedures in patients with Chiari malformation.

The BAEP changes seen during surgery can be divided into three types:

- **Type 1:** Gradual and persistent prolongation of the waveforms of 1 ms or more. Postoperative type 1 BAEP abnormalities are not accompanied by clinically significant hearing deficits, but careful audiological testing may reveal some minor hearing loss.
- **Type 2:** When there is a sudden loss of wave I through wave V ipsilateral to the side of the surgery without return to the baseline. Hearing impairment is often observed postoperatively on the same side of the surgery when type 2 changes are seen during surgery.
- **Type 3:** When the contralateral BAEP waveforms become abnormal during posterior fossa surgery, the prognosis is poor. Type 3 changes are often associated with other signs of brainstem dysfunction.

VISUAL EVOKED POTENTIALS

- The visual evoked potential (VEP) is an evoked electrophysiological potential that can be extracted using signal averaging from the electroencephalographic activity recorded at the scalp.
- The VEP can provide important diagnostic information regarding the functional integrity of the visual system.
- Elicited by monocular stimulation with a reversing checkerboard pattern, they are recorded from the occipital region in the midline and on either side of the scalp.
- The VEP peak latency, amplitude and waveform are age-dependent.

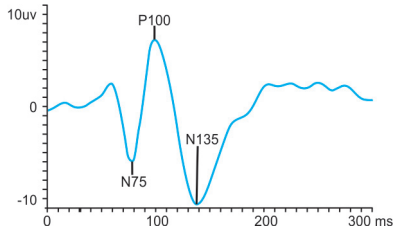


Fig. 3: Normal pattern of reversal visual evoked potential

- The VEP peak latency refers to the term implicit time used to describe the time from the stimulus to the maximum deflection of electroretinograms. The VEP peak latency may also be referred to as 'time to peak' or 'peak time'.
- The component of major clinical importance is the so-called P100 response, a positive peak having a latency of approximately 100 ms. (Fig. 3).
- The VEPs are most useful in detecting dysfunction of the visual pathways anterior to the optic chiasm.
- In patients with acute severe optic neuritis, the P100 is frequently lost or grossly attenuated.
- The VEP findings are, therefore, helpful in indicating previous or subclinical optic neuritis.
- They may also be abnormal with ocular abnormalities and with other causes of optic nerve disease such as ischaemia or compression by a tumour.
- Normal VEPs may be elicited by flash stimuli in patients with cortical blindness.

CARPAL TUNNEL SYNDROME

Aim of nerve conduction study (NCS) and EMG:

- To demonstrate a distal lesion of the median nerve.
- To exclude other peripheral conditions that can result in similar symptoms like high median neuropathy, C6-C7 radiculopathy, lesions of the brachial plexus or even polyneuropathy.
- To assess severity of carpal tunnel syndrome (CTS) and for therapeutic decisions.
- Baseline to assess the outcome after intervention.

Electrodiagnostic Grading of Carpal Tunnel Syndrome

Grade 1: Very mild CTS—normal standard tests, abnormal comparative tests.
Grade 2: Mild CTS—abnormal sensory with a normal motor response.
Grade 3: Moderate CTS—abnormal median sensory and motor response.
Grade 4: Severe CTS—absence of sensory response, abnormal distal motor latency.
Grade 5: Extreme CTS—absence of median motor and sensory responses.

Electrophysiological Findings

Sensory Latency

- Most important, most sensitive and earliest indicator of CTS is prolonged sensory latency.
- May show diminished amplitude and is often absent.
- Latencies of 1–2 ms are considered mild, whereas latencies of more than 6 ms are considered severe.

Distal Motor Latency

- Prolonged, but is not as sensitive an indicator as sensory latency.
- Motor latency abnormalities tend to occur later in the course of the disease.
- Latencies of 1–2 ms are considered mild, whereas latencies of more than 6 ms are considered severe.

Postoperative Changes

- There is an immediate increase in motor conduction velocity following release of the carpal tunnel.
- After one week, this value decreases to an intermediate value and then gradually returns to normal in the next 8–12 weeks.

Criteria for the Electrodiagnostic Evaluation of Unilateral Neurogenic Thoracic Outlet Syndrome

All three of the following criteria must be found in the affected limb:

- Amplitude of median motor response is reduced.
- Amplitude of ulnar sensory response is reduced.
- EMG evidence of denervation in muscles innervated by the lower trunk of the brachial plexus.

ACOUSTIC NEUROMA

- The results of pre-operative brainstem auditory-evoked response (BAER) studies were useful in predicting the outcome of hearing preservation attempts.
- Patients with intact BAER waveform morphology and normal or delayed latencies had a higher probability of hearing preservation in comparison to those with abnormal pre-operative BAER morphology.
- A characteristic finding on ABR in a person with an acoustic neuroma would be a wave I with nothing after it—no waves 3 or 5 (10–20% of cases). A wave I–III interval delay is common, and a wave V delay occurs in 40–60% of cases.

ELECTROENCEPHALOGRAPHY

- The electroencephalogram (EEG) was developed by the German psychiatrist, Hans Berger, in 1929.
- The EEG continues to play a central role in the diagnosis and management of patients with seizure disorders.
- The electroencephalograph records spontaneous electrical activity generated in the cerebral cortex.
- This activity reflects the electrical currents that flow in the extracellular spaces of the brain, and these reflect the summated effects of innumerable excitatory and inhibitory synaptic potentials upon cortical neurons.
- This spontaneous activity of cortical neurons is influenced and synchronised by subcortical structures, particularly the thalamus and upper brainstem reticular formation.
- Afferent impulses from these deep structures are probably responsible for entraining cortical neurons to produce characteristic rhythmic brain wave patterns such as alpha rhythm and sleep spindles.
- The EEG provides confirmation of Hughlings Jackson's concept of epilepsy—that it represents a recurrent, sudden, excessive discharge of cortical neurons; but like other ancillary tests, it must be used in conjunction with clinical data.

Applications of EEG

Diagnosis of Epilepsy

- Differential diagnosis of paroxysmal neurological events.
- Distinction between a focal and generalised seizure disorder.
- Identification of specific epilepsy syndromes.
- Recognition of photosensitivity and reflex epilepsies.

Management of Epilepsy

- Assessing risk of recurrence after an unprovoked seizure.
- Selection of antiepileptic treatment.
- Likelihood of seizure relapse if medication is withdrawn.
- Identification of irritative zone in epilepsy surgery candidates.
- Investigation of cognitive decline, especially when rapidly progressive.
- Monitoring in the management of status epilepticus.
- Detection of non-convulsive status.

Indications for Video-electroencephalography Monitoring

- Identification of epileptic paroxysmal electrographic and/or behavioural abnormalities.
- Verification of the epileptic nature of the new "spells" in a patient with previously documented and controlled seizures.
- Classification of clinical seizure type(s) in a patient with documented but poorly characterised epilepsy.
- Characterisation (lateralisation, localisation, distribution) of EEG abnormalities, both ictal and interictal, associated with seizure disorders.
- Quantification of the number or frequency of seizures and/or interictal discharges and their relationship to naturally occurring events or cycles. Quantitative documentation of the EEG response (ictal and interictal) to a therapeutic intervention or modification (e.g. drug alteration).

Partial Seizures

- Partial seizures, in scalp EEGs, are metamorphic, i.e. they show two or more distinct phases.
- The most common patterns consist of a series of rhythmic waves, sequential spikes/sharp waves, a mixture of spikes and rhythmic waves or regional voltage attenuation.
- Most often the initial frequency of temporal lobe seizures is in the alpha or theta range with slower frequencies occurring in a lesser proportion.
- Extratemporal seizures, however, often start in the beta frequencies rather than slower frequencies.
- Focal electrodecremental events are of excellent localising value, reflecting intense neuronal depolarisation or high-frequency firing.
- It is important to recognise that simple partial seizures, especially those with sensory rather than motor symptoms, may not be associated with discernable changes in routine scalp EEG in up to 80% of patients.

Generalised Seizures

- Typical absence seizures are characterised by isomorphic and stereotyped patterns that do not evolve as partial seizures. However, the spike-wave discharges may change from 3.5 or 4 Hz at the onset to 2 or 3 Hz as the seizure progresses. Also, the spike amplitude may decrease during the later part of the seizure.

- Atypical absence attacks frequently show gradual onset and offset with spike-wave discharges occurring at frequencies less than 3 Hz.
- Generalised tonic-clonic seizures may be preceded by diffuse polyspike-wave complexes. Ictal recordings during the tonic phase typically show generalised attenuation with or without high-frequency rhythmic waves that gradually increase in voltage ("epileptic recruiting rhythm") and evolve into polyspikes
- Myoclonic seizures are associated with 10–15 Hz polyspikes with or without slow waves, whereas tonic seizures show generalised paroxysmal fast activity or diffuse voltage attenuation preceded or followed by sharp and slow wave complexes.
- Generalised atonic seizures may show 2–3 Hz spike-wave discharges or may not be associated with any scalp EEG change.

ELECTROCORTICOGRAPHY

- This is a technique of placing grids or strips directly on the brain for recording EEG activity to localise the epileptogenic focus during surgery for epilepsy.
- Electrocorticography (ECoG) along with cortical stimulation mapping was first described by Penfield in 1939.
- The hypothesis for successful epilepsy surgery is based on the principle that removal of both lesional as well as the surrounding epileptogenic area is necessary for achieving seizure freedom. For greater precision, either intraoperative or extraoperative, subdural ECoG is often used to guide surgical resection of both the lesion and the epileptogenic zone.
- The area of interictal spiking or the irritative zone is often wider than the ictal onset zone (area where the seizure originates) which is considered as a gold standard for localising the epileptogenic zone.
- Intraoperative ECoG is widely utilised for electrical mapping of the epileptogenic zone during epilepsy surgery. It is useful to delineate the margins of the epileptogenic zone, guides the surgeon in achieving resection and is also of value to evaluate the completeness of resection.
- It has been found to be particularly useful in resective surgeries of neocortical foci (especially developmental lesions like cortical dysplasias) and for tailored resections in hippocampal sclerosis. The ECoG can be a valuable tool during multiple subpial transections (MST).

The obvious advantages of intraoperative ECoG are:

- They allow placement of recording and stimulation electrodes.
- Recordings can be performed before and after each stage of resection to assess the completeness of surgery.
- It allows direct electrical stimulation of the brain so that the regions involved in functions may be spared by the resection (e.g. eloquent cortex).
- No risks associated with long-term placement, e.g. infection.

The major limitations of ECoG are:

- The limited sampling time.
- Spontaneous epileptiform activity consists exclusively of interictal spikes and sharp waves, and seizures are rarely recorded. Thus, in most of the cases, localisation of the ictal focus is based on a hypothesis that it corresponds to the interictal activity, which is yet to be proven.
- It is difficult to distinguish primary ED from secondarily propagated discharges arising at a distant epileptogenic site.
- Both the background activity and the ED may be altered by the anaesthetics, narcotic analgesics and by the surgery itself.

CHAPTER 2

Intracranial Pressure

Ravi Ramamurthi • Nigel Peter Symss

HISTORY

- Quincke, in 1891, first reported the measurement of intracranial pressure through the lumbar route.
- Later studies by Quickenstedt, Ayala and Ayer established the range of normal ICP and demonstrated the effect of changes in body position and respiration, especially the Valsalva manoeuvre.
- Lundberg, in 1960, published his classic monograph on the continuous recording of ICP using an indwelling intraventricular catheter in a large series of 130 neurosurgical patients. He described three waveforms and sought to correlate the clinical features with changes in the pressure wave pattern.
- However, it was not before the 1970s that ICP monitoring came to be routinely used in clinical neurosurgery (Fig. 1).

ANATOMY AND PHYSIOLOGY

- The cranium can be likened to a rigid sphere. The three main components of this sphere are brain, blood and cerebrospinal fluid (CSF) occupying 1400 mL, 75 mL and 75 mL of space, respectively. The contents of the intracranial space are bound by the dura and communicate with the spinal canal through the foramen magnum.
- Since the thick bones of the calvarium are essentially non-distensible, the volume of the intracranial space is virtually constant regardless of the pressure generated within it. Therefore, any change in the volume of the brain causes a reciprocal change in the volume of other intracranial components, i.e. either blood or CSF. This is the basis of the modified

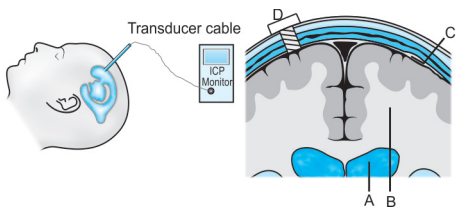


Fig. 1: Diagrammatic representation of ICP monitoring. Figure showing various methods of monitoring the ICP. (A) Intraventricular catheters. (B) Intraparenchymal catheters. (C) Epidural catheters. (D) Subarachnoid bolt

Monro-Kellie doctrine introduced into neurosurgery by Cushing. However, the Monro-Kellie doctrine does not hold true in infants because the skull is not rigid.

- The normal ICP is pulsatile due to the respiratory and cardiac cycle. The cardiac pulsations are reflected in the ICP through pulsations of the choroid plexus, and the cerebral and spinal arteries.
- The variations in venous return and cardiac output with respiratory excursions possibly account for changes in the ICP with the respiratory cycle.
- The normal CSF pressure measured through the lumbar route ranges from 50 to 200 mm of H_2O in the lateral decubitus position. The amplitude of the pressure wave can be as much as 5 cm of H_2O due to the combined effect of the cardiac and respiratory cycles.
- As the ICP increases, the pulse pressure generally increases.
- Lundberg described three pressure waves namely: A waves; B waves and C waves (Fig. 2).

A Waves

- A waves are pathological, there is a rapid rise in ICP up to 50–100 mmHg followed by a variable period during which the ICP remains elevated followed by a rapid fall to the baseline.
- The A waves that persist for longer periods (usually 5–20 minutes) are called plateau waves.
- Smaller A waves termed "atypical" or "truncated" A waves, that often do not exceed an elevation of 50 mmHg, are also clinically important early indicators of neurological deterioration.
- The A waves are accompanied by clinical features of raised ICP, e.g. headache, vomiting, decerebrate posturing, pupillary changes, bradycardia and hypertension, and respond to CSF drainage, hyperventilation and osmotic diuretics.

B Waves

- Occur at the rate of 0.2–2 per minute and are related to respiration.
- B waves may be vasomotor in origin. Lundberg initially described them in patients with intracranial hypertension, though they can also occur in normal individuals.
- B waves are said to be one of the best predictors of outcome after surgery for normal pressure hydrocephalus.

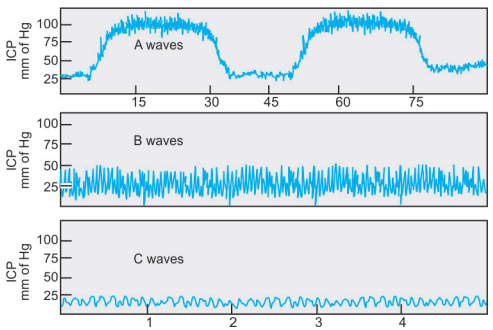


Fig. 2: Lundberg's CSF pressure waves

C Waves

- These are of low amplitude with a frequency of 4–8 per minute. These waves are thought to be related to Traube-Hering-Mayer waves.
- These waves are of little clinical significance.
- Increased ICP is indicated by a sustained elevation in pressure above 15 mmHg or when intermittent A or B waves are recorded.

VOLUME-PRESSURE RELATIONSHIP

- The cranium being rigid and non-distensible, any increase in the volume of a component would be accompanied by a reciprocal decrease in the volume of the other two components. Once the volume buffering capacity is exhausted, the ICP would begin to rise.
- During gradual expansion of a mass lesion, the volume displaced may be CSF, intravascular blood or brain tissue water. Of the three components, CSF appears to be the main buffer and is the first to be displaced as evidenced by compressed ventricles and obliterated subarachnoid spaces.
- The rate of expansion of an intracranial mass is also important. A rapidly growing intracranial mass lesion may outpace the compensatory shift of CSF and then even the smallest increase in mass could produce a life-threatening increase in ICP. Thus, a large haematoma could be accommodated within a few hours without a dangerous rise in ICP.
- Langfitt et al. studied the volume-pressure relationship in the rhesus monkey (Fig. 3). The flat portion of the curve was termed the period of spatial compensation, and the vertical portion was called the period of spatial decompensation. The curve may shift to the left if the intracranial mass expands very rapidly or if there is a pre-existing pathology in the brain reducing the amount of displaceable volume. The classical pressure-volume curve is exponential. In a semi-logarithmic co-ordinate system, the curve would be linear.
- Various mathematical models have been described to define the volume-pressure relationship (Fig. 4).
- **Compliance** is defined as change in volume per unit change in pressure (dV/dP). It is a measure of the distensibility of the intracranial space. The higher the compliance, the larger is the extra volume that the cranium can accommodate without a precipitous rise in pressure.

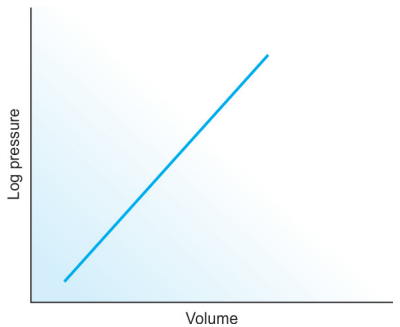


Fig. 3: The lineal relationship between the volume and the pressure semi-logarithmic co-ordinate systems

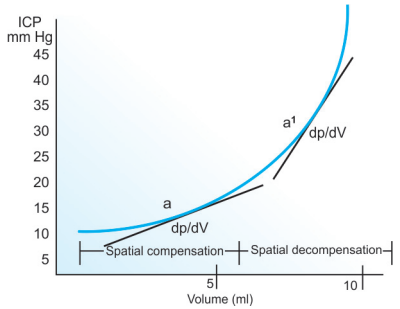


Fig. 4: Intracranial volume-pressure relationship. Note that the elastance at point 'a^1' is more than at point 'a'

- **Elastance** is inversely related to compliance (dP/dV). It is a measure of the resistance offered to an expanding intracranial mass. The slope of the volume-pressure curve is the elastance.
- In the horizontal limb of the volume-pressure curve, compliance is high and the elastance is low, while in the vertical limb, compliance is low and the elastance is high.

PATHOLOGY OF INCREASED INTRACRANIAL PRESSURE

- Intracranial hypertension can lead to secondary changes by interfering with the cerebral blood flow (CBF) and by producing brain herniation and pressure on critical structures.
- The average normal CBF is 50 mL/100 g/min. Ischaemic changes can develop when the CBF drops to 20 mL/100 g/min.
- The ICP influences the CBF through changes in the cerebral perfusion pressure (CPP).
- The CPP is defined as the difference between mean arterial pressure (MAP) and ICP, i.e. CPP = MAP – ICP. A rise in ICP would lead to a fall in CPP unless buffered by a rise in MAP.
- ICP and CPP monitoring are important in the management of head injury patients. However, there are other brain-related measures defined as 'pressures', such as cerebral intratissue pressure, critical closing pressure, 'optimal' CPP, non-invasive CPP (nCPP) and noninvasive ICP (nICP), and interhemispherical pressure gradients, which currently draw more attention in the management of head-injured patients.
- Raised ICP can cause arterial hypertension (the Cushing response), bradycardia and respiratory changes.

Intracranial Pressure Monitoring

- ICP monitoring has extensively been used in patients with head injury. It is most useful in neurologically stable (GCS 9 or more) patients with a traumatic intracranial haematoma, in whom the decision to surgically evacuate the haematoma is equivocal. A patient with persistently high ICP or a progressively rising ICP not responding to decongestants would merit surgical intervention.

- The current Brain Trauma Foundation recommendation of ICP monitoring in those patients presenting with a GCS score <8 with an abnormal CT scan or a normal CT scan with age >40 years, systolic blood pressure <90 mmHg or exhibiting posturing should be followed.
- Some neurosurgical centres monitor ICP in patients with aneurysmal subarachnoid haemorrhage (SAH).
- Continuous ICP monitoring in patients with suspected arrested hydrocephalus and normal pressure hydrocephalus can help to choose patients who may benefit from a CSF diversionary procedure.
- Neurosurgical patients who are paralysed and are being electively ventilated in the postoperative period should undergo ICP monitoring. Even in patients who are not being artificially ventilated, postoperative ICP monitoring can be beneficial if the haemostasis was difficult or the brain was not lax at the end of surgery.
- **Contraindications** to ICP monitoring include widespread scalp infection, any intracranial infection, open compound injury and bleeding diathesis.

Monitoring Systems

Monitoring systems can be divided into:

Fluid coupled system: It involves a fluid-filled catheter or a hollow bolt placed in the ventricle, subarachnoid space or the subdural space, connected to a pressure transducer through a fluid-filled line. The transducer converts the hydraulic pressure into an electrical signal which can be displayed digitally or on an oscilloscope.

Non-fluid coupled systems: In this, the transducer is mounted on the monitoring device itself. The monitoring devices have variously been designed for use in the ventricle, brain parenchyma, subdural and epidural space. This design has the advantage of being solid state and can be used in the ventricle, parenchyma or in the subdural space.

The telemetric system consists of an implanted transducer which is telemetered through the intact scalp. This system should particularly be useful for long-term ambulatory monitoring.

1. **Epidural monitoring:** This is performed by placing the device in the epidural space via a burr hole. These are simple to insert with a low infection rate. It is relatively safe in conditions, like fulminant hepatic failure, where haemostatic abnormalities may coexist and insertion of a parenchymal or ventricular device could be dangerous.
2. **Subarachnoid and subdural devices:** The subarachnoid and subdural devices are similar. The most commonly used devices for fluid coupled subdural pressure monitoring are the hollow screw or bolt devices, the "subdural cap catheter" and simple catheters placed in the subdural space. Intracranial pressure can also be measured from a subarachnoid catheter.

   The use of the Camino fibreoptic subdural device for the measurement of ICP in patients has been found comparable with recordings from the intraventricular fluid-filled catheter.
3. **Parenchymal monitoring:** This has become popular in some centres since the availability of non-fluid-filled devices, like the fibreoptic system and the piezoresistive microtransducers which can easily and safely be implanted into the brain parenchyma.

Parenchymal monitoring has been useful in patients with small ventricles or large shifts, in large craniectomies or open compound injuries with dural loss where placement of an epidural or subdural device may be difficult.

4. **Intraventricular monitoring:** This remains one of the most popular techniques and is probably the procedure of choice in patients with ventriculomegaly.

- It is very accurate, and is the current gold standard against which other methods are compared.
- A variety of sets are available commercially, but basically they all consist of a ventricular catheter which is fluid coupled to a strain gauge transducer.
- The placement of the catheter is simple and the standard landmarks for a ventricular tap are followed. It is usually placed in the frontal horn and tunnelled from its site of exit from the skull to the point of exit from the scalp. Placement of the catheter is verified by the drainage of fluid and an adequate waveform on the monitor.
- It offers the added advantage of therapeutic drainage of CSF to lower the ICP, and can also be used to perform CSF dynamic studies.
- The major disadvantage is that it may be difficult to insert in patients with small ventricles or significant shifts. The risk of infection varies from 8% at 5 days to 40% at 12 days. Intracranial hemorrhage can occur. The failure rate is considerably lower than with the subarachnoid bolt.
- ICP monitoring from the operative cavity in the posterior fossa using a fluid-filled catheter is safer and more accurate than supratentorial monitoring after posterior fossa surgery. It also has the added advantage of allowing drainage of CSF or collected blood whenever required.

CHAPTER 3

Neuro-ophthalmology

Tandon R • Saxena R • Phuljhele S

EFFECTS OF BRAIN TUMOURS ON VISUAL PATHWAYS

Direct effects:

- Direct compression.

Indirect effects: Papilloedema

- Impact of tumour resection
- Post-radiation neuropathy
- Toxic effects due to chemotherapy
- Metastatic lesions to retina or infiltration of optic nerve
- Haematological effects like ischemia, venous engorgement.

THE NORMAL FIELD OF VISION

- The visual field is a three-dimensional area which is visible through each eye when fixating at a central target.
- The field of vision is centred at a fixation point of each eye, as an asymmetric oval shape extending maximally on the temporal side for about 90 degrees.
- The superior and inferior fields have an extension of 40 and 60 degrees, respectively. Nasally it is restricted by the nasal bridge and measures up to 40 degrees.
- There is binocular representation of the visual fields for 60 degrees to the right and left of a common fixation point.

THE NORMAL VISUAL PATHWAY

- The visual pathway begins with stimulation of photoreceptors of the retina and processing in the ganglion cells. The axons of the ganglion cells converge to form the optic nerve.
- The nerve then travels through the optic canal and emerges in the intracranial cavity at the sellar region. At this juncture, the nasal fibres of the optic nerve cross to form the optic chiasm. The contralateral nasal fibres and ipsilateral temporal fibres continue together as the optic tract, which provides for one-half of the binocular visual field.
- The optic tract is relayed in the ipsilateral lateral geniculate body (LGB) where sorting of fibres occurs.
- The third order neurons commence from the LGB and continue as the optic radiation that terminates in the visual cortex (area 17) of the occipital lobe (Fig. 1).

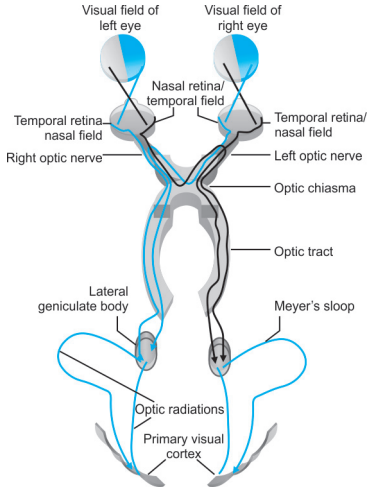


Fig. 1: Visual pathway

ASSESSMENT OF VISUAL FIELD

- The process of estimating and mapping the field of vision is called perimetry. The field of vision of a patient can be assessed by various methods; the easiest and convenient for bed-side examination is confrontation examination. This is a crude and subjective evaluation, wherein the examiner compares the visual field of the patient to his own field of vision.
- More sophisticated methods used routinely in clinical practice include Goldmann perimetry and automated perimetry, which are objective and, hence, more reliable methods. These techniques can precisely map and record the peripheral (full extent) as well as central (central 30°) field of vision, respectively.

INTERPRETATION OF VISUAL FIELD

Terminologies

Isopter: It is a line joining the retinal points of the same sensitivity.

Constriction: It implies that there is generalised decrease in sensitivity of the retina and, on kinetic perimetry, it encloses a smaller area than normal.

Hemianopia: When one-half of the visual field is involved either in one or both the eyes.

Homonymous: When the defects are present in the visual fields of both the eyes and are on the same side of the vertical meridian.

Heteronymous: When the defects are present in the visual fields of both the eyes but are on the opposite sides of the vertical meridian.

Scotoma: It is a localised area of decreased sensitivity. It may be absolute, when the patient cannot see even the brightest possible stimulus or it may be relative if some of the stimuli are seen. They can be classified either by their location or by shape.

Congruency: The extent to which the homonymous field defect in one eye resembles that in the other eye in terms of shape, size, depth and slope of the margins.

Table 1: Visual field defects according to the location of the tumour

Location of tumours	*Structure involved*	*Type of field defect*
Orbit		
• Optic nerve glioma • Optic nerve meningioma	• Optic nerve	Total field loss (early lesions may present with central or centrocecal field defect)
Sella		
• Pituitary adenoma • Craniopharyngioma • Meningioma	• Chiasma • Chiasma • Willebrand's knee • Optic tract	• Bitemporal field defect • Bitemporal field defect • Junctional scotoma • Homonymous hemianopia (macular splitting)
Mid brain		
• Craniopharyngioma • Meningioma • Large pituitary adenoma • Aneurysms • Hamartomas	• Lateral geniculate body	• Sectoral defects
Parietal lobe		
• Meningiomas • Gliomas • Metastasis • Vascular	• Superior part of optic radiation	• Inferior quadrantic field defect • Homonymous hemianopia (macular splitting)
Temporal lobe		
• Gliomas • Metastasis • Vascular	• Inferior part of optic radiation	• Superior quadrantic field defect (usually incongruous in nature)
Visual cortex		
• Gliomas • Meningiomas • Metastasis • Vascular	• Occipital lobe	• Homonymous hemianopia sparing macula. (Complete or partial depending upon the size of the tumour)

VISUAL FIELD DEFECTS

- The arrangement of visual fibres in a particular region of the visual pathway determines the characteristics and shape of visual field defects.
- In general, lesions anterior to the optic chiasma produce monocular defects, while lesions affecting the chiasma and beyond are always bilateral.
- Heteronymous defects are seen in chiasmal lesions, while all lesions affecting the visual pathway beyond the chiasma produce homonymous lesions.
- The more posterior a lesion is in the visual pathway, the more congruent field defect it will produce. For example, a lesion of the occipital lobe will produce a more congruent field defect than that of the optic tract.

Retina

- The visual field and the retina have an inverted and reversed relationship. Thus, light rays from the temporal visual field stimulate the ganglion cells of the nasal retina. The axons of these ganglion cells form the nerve fibre layer and finally converge at the optic nerve head and exit as the optic nerve.
- The innermost layer of the retina, retinal nerve fibre layer (RNFL) bundles converge in a specific pattern to form the optic nerve head.
- The macular fibres are centrally placed, while fibres from the upper part of the retina lie superiorly and those from the lower retina are placed inferiorly in the optic nerve.

- The layer of the retina that is affected by the disease process determines the pattern of field defect. Lesions in the outer retina have no particular definite boundaries, so they can produce field defects of varying shapes and sizes, while involvement of the RNFL produces a field defect corresponding to the particular pattern of the nerve fibre bundle affected.
- ***Papillomacular bundle*** involvement results in a central scotoma, centrocecal scotoma or paracentral scotoma.
- ***Arcuate nerve fibre bundle*** lesions may cause a variety of field defects like—Bjerrum or arcuate scotoma, Seidel scotoma, Nasal step of Ronne, depending on the site of the lesion. Arcuate field defect is seen in many conditions, apart from anterior optic nerve lesions (see the following box):

- Glaucoma
- Chorioretinal lesions
- Juxtapapillary choroiditis
- Myopia with peripapillary atrophy
- Central retinal artery occlusion
- Branch retinal artery occlusion
- Ophthalmic artery or carotid artery occlusion
- Drusen
- Optic nerve pit
- Chronic papilloedema
- Anterior optic nerve lesions
- Involvement of the nasal fibre bundle leads to temporal wedge-shaped defects.

Optic Nerve

- The optic nerve is formed by the axons of retinal ganglion cells that converge to form the nerve fibre bundles. It is about 50 mm in length and can be divided into four parts: optic nerve head (intraocular part 1 mm), intraorbital (25 mm), intracanalicular (10 mm) and intracranial (15 mm).
- The above described arrangement of the nerve fibres is maintained throughout the optic nerve, such that even lesions involving the posterior part of the optic nerve produce a pattern corresponding to the nerve fibre bundle involved (Fig. 2).
- In addition to the field defects described above, the involvement of axons at the superior or inferior pole of the optic disc produce *altitudinal defects*.

The Chiasma

- It is formed by the crossing of the nasal fibres of each optic nerve. It is located over the diaphragm sella and is posteriorly related to the wall of the third ventricle.
- In 80% located directly over the sella (central); in such cases, pituitary lesions involve the chiasma first. In about 10%, it is located more anteriorly over the tuberculum sellae (prefixed); here pituitary lesions will involve the optic tract first. In the remaining 10% of cases, it is located more posteriorly over the dorsum sella, where pituitary lesions involve the optic nerve first.

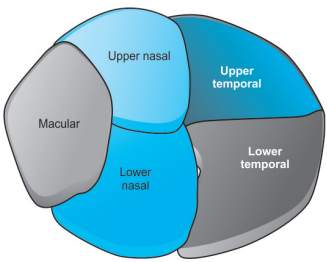


Fig. 2: Arrangement of nerve fibres in optic nerve

- **Anteriorly**, the chiasma is related to the anterior cerebral arteries and their communicating branches. **Posteriorly**, it is related to the hypophyseal stalk and the pituitary body. It is immediately inferior to the floor of the third ventricle and lies above the hypophysis. **Laterally**, it is related to the internal carotid artery and the cavernous sinus.
- At the chiasma, the fibres representing the nasal visual field (temporal fibres) are present in the lateral part. The inferonasal fibres (superior temporal field) cross anteriorly in the chiasma and loop forward into the contralateral optic nerve, forming von Willebrand's knee and then continue in the contralateral optic tract. The nasal macular fibres decussate most posteriorly. The upper nasal fibres (inferior temporal field) cross in the middle (Fig. 3). Thus, the right optic tract constitutes the right nasal field (right temporal axons) and the left temporal field (left nasal decussated fibres) representing the left half of the visual space.
- A patient presents with varying signs and symptoms, depending upon the type, size and site of the lesion. Pituitary adenoma presents with signs of compression of the visual pathways, as well as other features of hormonal disturbance. There may be unilateral or bilateral loss of vision, as a result of concurrent compression of the optic nerve by the tumour. Relative afferent pupillary defect may be present in unilateral lesions. Lesions of the cavernous sinus and large pituitary adenomas compress the motor nerves of the eye, leading to ocular motility disorders. See-saw nystagmus and oscillopsia are features of tumour or trauma.
- **Field Defects:**
  - ***Bitemporal hemianopia:*** Lesions that affect the body of the chiasma produce bitemporal hemianopia. Lesions of the pituitary gland, i.e. tumours, inflammation or haemorrhage are the common conditions. Inferior bitemporal hemianopia is characteristic of craniopharyngioma. Saccular aneurysms of vessels in the circle of Willis can also lead to bitemporal hemianopia.
  - ***Junctional scotoma:*** It is produced when a lesion involves the junction of the optic nerve and chiasma, leading to an optic nerve defect in the ipsilateral eye and superior temporal (involvement of von Willebrand's knee) defect in the contralateral eye.
  - ***Binasal field defects:*** A large mass lesion compressing one side of the optic chiasma, on further enlargement, causes a midline shift of the chiasma with subsequent compression of the other side as well, leading to a binasal field loss presentation. This pattern of field loss

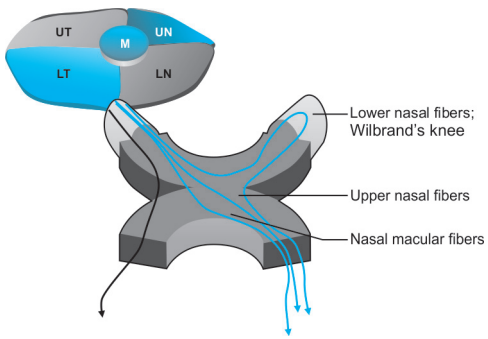


Fig. 3: Decussation of optic nerve fibres at the chiasma

can also be seen with empty sella syndrome (ESSD), arachnoiditis and after surgery for pituitary tumours.
- ***Homonymous hemianopia:*** This type of field defect is commonly seen in cases where the chiasma is prefixed. Thus, any lesion which affects the chiasma can also affect the optic tract, leading to homonymous hemianopia. Large pituitary adenomas and craniopharyngiomas are the most common causes.
- Lesions that affect the posterior part of the chiasma cause *central bitemporal hemianopia*.

Optic Tract and Lateral Geniculate Body

- The optic tract runs superiorly and posteriorly from the optic chiasma and curves around the brainstem to terminate in the LGB. The LGB is a part of the thalamus and is situated along the lateral aspect of the midbrain. Each geniculate body consists of six layers of neurons and second order fibres, travelling through the optic tract, are relayed in different layers of the LGB. It receives dual blood supply from the anterior and lateral choroidal arteries, which nourish the fibres from the superior and the inferior homonymous quadrants of the retina, respectively. The optic tracts lie between the tuber cinereum and the anterior perforated substance.
- Each optic tract is formed by the temporal fibres of the ipsilateral eye and the nasal fibres from the contralateral eye.
- The axons from the ipsilateral eye are relayed in nuclei layers 2, 3 and 5, while the axons from the contralateral side relay in 1, 4 and 6 nuclei layer. The fibres from the upper part of the retina occupy the medial half of the anterior one-third of the LGB. The lower retinal fibres occupy the lateral half of the anterior one-third of the LGB, while the macular fibres are relayed in the posterior two-third of the LGB.
- The most useful sign of localising a lesion between the optic tract and LGB is involvement of the pupil. Optic tract lesions are associated with various pupillary abnormalities including RAPD on the contralateral side, Wernicke's hemianopic pupil and Behr's pupil, while in lesions of the LGB, pupillary reactions are spared.
- Lesions affecting the optic tracts are craniopharyngiomas, meningiomas, and large pituitary adenomas, demyelinating diseases, vascular lesions, such as aneurysms, arteriovenous malformations and hamartomas.
- Lesions affecting the LGB include infarction, tumours, trauma and inflammatory disorders.
- Complete optic tract disruption on one side leads to complete macular splitting homonymous hemianopia, while partial involvement will cause an incongruous field defect. Similarly, a defect in one LGB will produce complete homonymous hemianopia, while partial lesions of the LGB cause a sectoral field defect.

Optic Radiations

- The optic radiations are geniculocalcarine pathways that extend from the LGB to the visual cortex. They cross the Wernicke's area as optic peduncles and travel in the retrolenticular part of the internal capsule.
- The fibres, after exiting from the LGB, rotate by 90 degrees again to regain their original arrangement. Thus, the superior fibres lie in the upper part of the radiation, the inferior fibres in the lower part and the macular fibres lie in the centre.

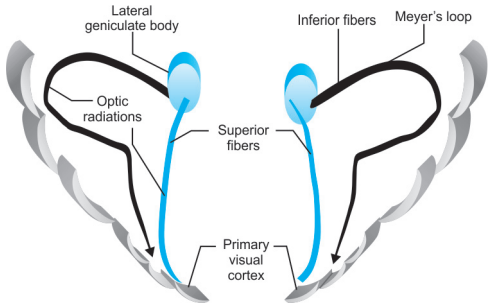


Fig. 4: Arrangement of optic radiations

- ***Arrangement of fibres in the temporal lobe:*** The inferior fibres from the corresponding superior visual fields of both eyes commence from the anterolateral part of the LGB, sweep anteriorly and then laterally along the temporal horn of the lateral ventricle to form Meyer's loop, then extend posteriorly to end in the area below the calcarine fissure of the occipital cortex (Fig. 4). The clinical features of temporal lobe lesions are seizures, paragnosia, involuntary movement of the mouth, and visual and auditory hallucinations. Aphasia is characteristic of left temporal lobe lesions. The most common temporal lobe lesions are tumours, which include glioma and metastasis. Surgical resection of the tumour (when more than 8 cm), itself may produce field defects. Anterior choroidal artery occlusion may affect the optic radiation in the temporal lobe and LGB. Other lesions are trauma, demyelination and abscess.
- ***Arrangement of fibres in the parietal lobe:*** The superior fibres that carry the inferior field travel directly through the lower portions of the parietal cortex to the occipital lobe area above the calcarine fissure (Fig. 4).

Clinical Features of Parietal Lobe Involvement

- The two most important localising signs of a parietal lobe lesion are loss of normal optokinetic response and conjugate deviation of the eyes. The involvement of the right parietal lobe leads to deficit of spatial orientation of the left space, thus the patient may neglect the left half of the body. Involvement of the left parietal lobe is associated with Gerstmann's syndrome (agraphia, acalculia and visual agnosia). The differential diagnosis of lesions involving the parietal lobe, include infarction, haemorrhage, AVM and tumours.
- Lesions involving the optic radiations produce field defects which are homonymous and more congruous. The involvement of the anterior temporal lobe will produce contralateral homonymous superior quadrantanopia. In more extensive lesions, the defect may extend inferiorly but the hemianopia will be denser superiorly. The defects are wedge shaped defects of varying sizes and are homonymous and incongruous.
- Lesions involving the parietal lobe produce an inferior homonymous field defect that is classically described as pie on the floor. These defects are usually more extensive and more congruous than temporal lobe defects. The homonymous hemianopia of the parietal lobe is always associated with macular splitting.

Visual Cortex

- The primary visual cortex (area 17) is located on the medial surface of the occipital lobe near the calcarine fissure. Brodmann's area 18 and

19 are associated visual areas which help in further processing of visual information.

- The right visual cortex receives impulses from the left temporal field and the right nasal field. The orientation of fibres in the optic radiation is maintained in the visual cortex. The central field is represented at the occipital tip and is separated from the representation of peripheral fibres. The superior fibres (representing inferior field) are relayed in the area superior to the calcarine fissure while the inferior fibres (superior field) are projected to the area inferior to the fissure.
- Occlusion of the left posterior cerebral artery causes infarction of the left calcarine cortex, along with the splenium of the corpus callosum, which produces field defects in the right side and alexia without agraphia. Other signs include headache, visual agnosia and dyschromatopsia, which are mainly secondary to the infarction. Tumours lead to isolated visual field defects and palinopsia. Tumours affecting the occipital lobe are gliomas, meningiomas and metastasis. Bilateral occipital disease is usually due to vascular lesions which include thromboembolism, haemorrhage, hyperviscosity syndromes and vasculitis. This is also a feature following bilateral tentorial herniation, which results in bilateral occipital infarct resulting in blindness.
- **Field Defects:**
  - Occipital lobe lesions produce field defects that are homonymous and highly congruous.
  - ***Complete homonymous hemianopia sparing the macula:*** The macular area is a watershed zone of anastomoses between the middle and the posterior cerebral arteries. Ischaemia of the occipital lobe results in sparing of the central 3–5 degrees of the visual field due to its dual blood supply.
  - ***Central homonymous hemianopia:*** A lesion affecting the tip of the occipital lobe would produce a field defect in the macular area which is homonymous. In such cases, at least 5 degrees of the central field is spared in both eyes on the side of hemianopia.
  - ***Quadrantic field defects:*** Lesions of the area below the calcarine fissure would give rise to a homonymous superior quadrantic defect. Similarly, a lesion involving the area above the calcarine fissure would lead to an inferior homonymous defect.

Involvement of the occipital lobe bilaterally may lead to *bilateral homonymous hemianopia* with sparing of central tubular vision. This should be differentiated from other causes of constricted visual field like malingering, glaucoma and retinitis pigmentosa.

SPECIAL VISUAL FIELD DEFECTS

Baring of Blind Spot

- When a small degree of field is mapped, the isopter may be constricted and any scotoma outside the isopter may be continuous with the blind spot causing baring of the blind spot.

Binasal Hemianopia

- They are bilateral nasal defects usually caused by involvement of arcuate fibres of both eyes. Rarely, caused by pressure upon the temporal aspect of the optic nerve or anterior portion of the chiasma, as in cases of aneurysm, pituitary tumour and vascular infarction.

4

CHAPTER

Neuro-otology

Sathiya Murali • Srividya • Mohan Kameswaran • Kiran Natarajan

QUANTITATIVE TESTS FOR VESTIBULAR FUNCTION

Electronystagmography

- Electronystagmography (ENG) remains the most useful laboratory test in the evaluation of patients with complaints of dizziness or vestibular disturbance.
- ENG can provide diagnostic information when there is a suspicion of unilateral or bilateral vestibular hypofunction.

Electro-oculography

- Electro-oculography (EOG) is typically used to record eye movements during ENG testing based on the corneo-retinal potential (electrical charge potential between cornea and retina).
- The eye acts as an electrical dipole along its long axis. Movement of this dipole relative to the surface electrodes produces an electrical signal corresponding to eye position. Horizontal eye movements can typically be resolved to an accuracy of 0.5°. The sensitivity of EOG is less than that of direct visual inspection (approximately 0.1°).
- Visual inspection of small amplitude eye movements directly or with Frenzel's lenses or an ophthalmoscope is important for documentation of low amplitude nystagmus.

Video-oculography/Videonystagmography

- Video-oculography (VOG) is infrared (IR) imaging analysis which uses the conventional black and white camera. The eyes are illuminated with IR light. Eyes are not reactive to IR light, and hence can be viewed while in total darkness, thus eye fixation is eliminated. The eye movements are recorded by an IR video camera and converted into a digital format through software that documents the eye movements.
- Horizontal and vertical tracings of eye movements are recorded by the camera tracking the pupil of the eye.
- Advantages of VOG include accuracy of 0.1–0.5°, contact free recording of eye movements and ease of handling. Rotatory eye movements in benign paroxysmal positional vertigo (BPPV) can be detected only in VOG.
- Static tests include spontaneous nystagmus and gaze nystagmus. Spontaneous nystagmus is suitable for recording non-evoked eye movements with eyes closed and with eyes open.

Gaze Nystagmus

- Here, the eye movements are measured while the patient is fixating on a target.
- Nystagmus caused by CNS lesions can be differentiated from that caused by peripheral vestibular lesions.
- CNS nystagmus may be horizontal, vertical or rotatory.
- If horizontal, CNS nystagmus usually beats to the right in rightward gaze and to the left on leftward gaze, and is usually not suppressed by ocular fixation. This is in contrast to nystagmus caused by a peripheral lesion. Here it is horizontal, always beats in one direction (usually toward the normal side and suppressed by ocular fixation).
- Nystagmus is named after the direction of the fast component which is the corrective movement of the eyes generated by the CNS. The vestibular system generates the slow phase of nystagmus which is directed to the opposite side (Table 1).

DYNAMIC TESTING

Saccade Testing

- The patient is instructed to fixate a series of randomly displayed dots or lights at eccentricities of 5–30° saccades.
- Saccades typically begin with a latency of 180–200 milliseconds after presentation of a target. Saccade velocity increases linearly with amplitude up to about 20° but remains relatively constant for higher amplitudes. Healthy subjects consistently undershoot the target for saccades more than 20°.
- Asymmetries in saccade amplitude or peak velocities can provide localising information.

Smooth Pursuit

- The patient is asked to watch a target that moves horizontally in a sinusoidal fashion at a low frequency with position amplitude of 20° in each direction.
- Inspection of the waveform of the tracking eye movement is often of greater diagnostic use than absolute measure of gain and phase.
- "Catch up" saccades are typically made when pursuit responses are decreased.
- Saccadic pursuit is characterised by the occurrence of these saccades in a "stair-step" pattern. Such patterns are seen in cerebellar disease and also may occur with decreases in pursuit gain that occur with ageing.

Table 1: Features differentiating central from peripheral causes of vertigo

Features	*Central*	*Peripheral*
Imbalance	Severe, mostly constant	Mild to moderate, mostly episodic
Neurologic symptoms	Frequent	Rare
Nystagmus	Changes direction in different gaze positions No change with visual fixation	Unidirectional in all gaze positions Decreases with visual fixation
Hearing loss	Rare	Frequent
Nausea	Variable, may be absent	Severe
Recovery by central compensation	Slow	Rapid

- Saccadic substitution of smooth pursuit is seen in neurological lesions.
- Disorganisation of pursuit eye movements with wandering slowed inaccurate tracking is seen in brainstem lesions.

Optokinetic Testing

- Testing is often performed with the subject surrounded by a visual scene that moves completely in one direction at velocities of 30–60° per second.
- The optokinetic tracing response is a nystagmus in the plane of motion of the visual scene.
- Asymmetries in OKAN have been reported in patients with unilateral vestibular hypofunction induced by removal of an acoustic neuroma and other CNS lesions. Responses are greater in amplitude and longer in duration for stimulus and eye movement toward the side of the lesion.

Caloric Testing

- Caloric testing remains the most useful laboratory test in determining the responsiveness of a labyrinth.
- It is one of the few tests that allow one labyrinth to be studied independently of the other.
- Caloric testing relies on stimulating or cooling the vestibular system by alternately heating and cooling the external auditory canal with water or air.
- The horizontal canal is affected most by such temperature effects, because it is located closest to the external auditory canal and is oriented in the plane of the temperature gradient that is produced in the temporal bone from irrigation with water.
- Calorics cause a response in two ways. The first is a convective component, with the temperature gradient across the horizontal canal resulting in a density difference within the endolymph of the canal. When the horizontal canal is oriented in the plane of gravity, the more dense fluid falls to the lower position in the canal, whereas the less dense fluid moves to the upper portion in the canal. In the presence of gravity, there is a flow of endolymph from the cooler (more dense) region to the warmer (less dense) region. This movement of fluid within the canal deflects the cupula, thereby leading to a change in discharge rate of vestibular nerve afferents. Endolymph flows toward the ampulla for warm irrigation and away from the ampulla for cold irrigation. This effect depends on head position.
- Alternate binaural caloric testing as pioneered by Fitzgerald and Hallpike is the most commonly used testing protocol. Cool water (30°C) and warm water (44°C) are administered for 60–90 seconds to each ear in a set order such as right warm, left warm, right cold and left cold. Such a stimulus results in heating effect in the temporal bone that lasts for 10–20 minutes.
- Cold caloric test has been utilized to evaluate brain stem function in comatose patients and for diagnosis of brain death.

Rotatory Chair

- Unlike caloric tests, rotational tests analyse the responses of both labyrinths together.
- Rotatory chair testing is useful in assessing vestibular function in patients with suspected bilateral vestibular hypofunction, in patients receiving vestibulotoxic medications and in children who may not tolerate caloric testing.

- Cerebellar abnormalities produce specific changes that can be evaluated with rotatory chair testing.

Vestibular Evoked Myogenic Potentials

- Vestibular evoked myogenic potentials (VEMP) is a sound-evoked muscle reflex, or sonomotor response that can be recorded using evoked potential techniques by acoustical stimulation of the saccule.
- VEMP has become an important investigative modality in the evaluation of patients with balance disorders.
- The responses are abolished on the side of surgery after unilateral vestibular neurectomy.
- VEMP may be absent in basilar artery migraine, Ménière's disease, idiopathic bilateral vestibulopathy (IBV) and vestibular schwannoma.
- VEMP may be increased in superior semicircular canal dehiscence syndrome and perilymphatic fistula.
- Asymmetrical amplitudes may be seen in Tullio's phenomenon and spasmodic torticollis. Delayed VEMP is seen in cases of technical error, central lesions like brainstem stroke, multiple sclerosis, spinocerebellar degeneration and migraine.

Computerised Dynamic Posturography

- Computerised dynamic posturography (CDP) is a series of vestibulo-spinal tests for quantitatively assessing balance function in different set-ups which simulate conditions encountered in our day-to-day life.
- CDP tests the two faculties of the balance system separately. The first portion is tested by the sensory organisation test (SOT) and the second portion by motor coordination test (MCT).
- The functional integrity of the sensory system (i.e. the inputs to the balance system, viz. the visual, proprioceptive and vestibular) is assessed by the SOT. This test detects any defect in the subject's ability to effectively use the somatosensory/visual/vestibular inputs to maintain balance and the ability of the CNS to select the appropriate input when contradictory and conflicting information is sent through these three input systems.
- The second part of the CDP test is the Motor Coordination Test or Motor Control Test (MCT). It is useful in assessing the integrity of efferent motor pathways in the control of balance.

Cranio-corpography

- Cranio-corpography (CCG) basically consists of photographically recording the patient's head and body movements as he or she performs the Unterberger's stepping test and the Romberg's test.
- CCG is a very quick, non-invasive and very simple objective test of vestibular and balance functions.
- It is totally physiological, easily repeatable and provides a photographic and quantifiable record, and can even be done in children and in patients with perforated ear drums.

INVESTIGATIONS FOR THE AUDITORY SYSTEM

- Puret one audiogram
- Impedance audiometry.

- Electrophysiological tests:
  - Otoacoustic emissions (OAE)
  - Electrocochleography (ECoG)
  - Auditory brainstem response (ABR)
  - Auditory steady state response (ASSR)
  - Middle latency responses (MLR)
  - Long latency response (LLR).

Pure Tone Audiogram

- Pure tone audiogram is a subjective test, which is used to detect if there is definite hearing loss, its type (sensorineural/conductive) and degree of the hearing loss.
- It helps in measurement of hearing threshold at various frequencies.
- The disadvantages of the test are that it is purely subjective, cannot be performed in a child, a patient who is not co-operative and in unconscious patients.

Impedance Audiometry

- Impedance audiometry has been one of the major advancements in the field of otology and neuro-otology in recent times.
- The uses of impedance can briefly be summarised as:
  - Objective differentiation between conductive and sensorineural hearing loss.
  - Measurement of middle ear pressure (tympanometry) and evaluation of Eustachian tube function.
  - Differential diagnosis of whether the lesion is cochlear or retrocochlear.
  - Identification of site of lesion in facial palsy and certain brainstem pathologies (stapedial reflex test).

ELECTROPHYSIOLOGICAL TESTS

Otoacoustic Emissions

- Otoacoustic emissions (OAE) are biological sounds from the normal cochlea. This sound is generated in the outer hair cells of the cochlea.
- This is another objective test for hearing screening in neonates and young children which has now become popular.

Electrocochleography

- Electrocochleography (ECoG) is a short latency evoked potential that reflects the summed activity of a large number of peripheral auditory nerve fibres as well as the response of generators located within the cochlea itself.
- The ECoG consists of three distinct evoked potentials:
  1. The cochlear microphonic (CM)
  2. The summating potential (SP)
  3. Compound action potential (AP)
- The CM and the SP are both intracellular potentials.
- The CM is a potential that mirrors the stimulus and it reflects the instantaneous displacement of the basilar membrane.
- The SP is characterised by a baseline shift in the CM and it is also thought to originate from within the hair cells of the organ of Corti.

- The AP is the third evoked potential of the ECoG complex. It is a recording of the synchronous response of a large number of auditory nerve fibres to the acoustic stimulation.
- The ECoG is recorded either extratympanically or intratympanically. It is rerecorded extratympanically via an active electrode placed within the ear canal or on the drum itself and a reference electrode placed on the contralateral mastoid process. Intratympanically, the ECoG is recorded using a transtympanic needle electrode placed on the promontory of the middle ear.

Clinical Applications

- To assess hearing in the paediatric age group.
- Tool to assess auditory status in patients suspected of having Ménière's disease.
- As a part of retrocochlear assessment.
- During otoneurologic surgery.

Auditory Brainstem Response

- Auditory brainstem response (ABR) is a recording of the synchronised response of a large number of neurons in the lower portions of the auditory pathways was first described by Sohmer and Feinmesser in 1967. Origin of various waves has been described in Table 2.
- The ABR is recorded using differential amplification with the active electrode positioned at the vertex or high forehead with reference electrodes positioned at the mastoids or the ear lobes.
- It requires brief acoustic stimulus with a relatively rapid onset. The most common stimulus used to evoke the ABR is a 100-millisecond rectangular pulse or click. A click is the ideal stimulus for eliciting the ABR as it results in a large number of auditory nerve fibres firing at approximately the same time.

Clinical Applications

- It is a tool for estimating auditory sensitivity as well as for a range of otoneurologic applications.
- It is by far the most commonly used of the auditory evoked potentials, due to the fact that it can be recorded by non-invasive techniques and is easy to record, not strongly affected by attention, sleep state, sedation and anaesthesia or age.
- Historically, the recording of ABR has been to assist with detection of lesions affecting the auditory pathways between the cochlea and the inferior colliculus. Such lesions like vestibular schwannomas and pathology, caused

Table 2: Origin of various waves of auditory brainstem response

Wave	*Site of origin*
I	Cochlear nerve (distal end)
II	Cochlear nerve (proximal end)
III	Cochlear nucleus
IV	Superior olivary complex
V	Lateral lemniscus/inferior colliculus
VI and VII	Neural generators, not definitely known

by factors such as multiple sclerosis, stroke or trauma.
- It is used to monitor the status of the auditory nerve during skull base surgery, because ABR is not strongly affected by anaesthesia. It provides continuous feedback about the status of the auditory nerve during surgery.

Auditory Steady State Response

Auditory steady state response (ASSR) is a far-field auditory potential that is evoked using continuous stimulation.

As a potential clinical tool, it has several advantages over the more commonly used ABR. For example, with ASSR, it is easier to distinguish between severe and profound hearing loss as opposed to ABR.

Vestibular Function Test in Unconscious Patients

Vestibulo-ocular reflex elicited by irrigating the external auditory canal with ice cold water has proved to be a very reliable bedside investigation to evaluate unconscious patients.

It not only provides objective indication of the depth of unconsciousness, evidence of ocular nerve palsies, internuclear ophthalmoplegia but is also a guide to prognosis.

COCHLEAR AND BRAINSTEM IMPLANTS

Mohan Kameswaran, Kiran Natarajan

- Auditory neural prostheses, such as cochlear implants and brainstem implants, have proved to be extremely useful innovations in neuro-otology in the last decade. This has been made possible due to advances in biomedical engineering and the development of materials that are biocompatible.
- Cochlear and auditory brainstem implants offer safe and effective hearing habilitation and rehabilitation for profoundly deafened adults and children.
- Auditory neural prosthetic intervention must be done as early as possible due to the phenomenon of neural plasticity. Neural plasticity is the ability of the central nervous system to be programmed to learn a new task. This fades between 6 years and 8 years of age.
- Electrophysiological tests and imaging modalities are now available to accurately pinpoint the level of lesion in the auditory pathway. In 99% of sensorineural hearing loss, including congenital hearing impairment, the primary pathology is in the cochlea.

COCHLEAR IMPLANTS

- A cochlear implant is a surgically implantable device that helps restore hearing in patients with severe or profound hearing loss, unresponsive to amplification by hearing aids. Cochlear implants are electronic devices designed to detect mechanical sound energy and convert it into electrical signals that can be delivered to the cochlear nerve, bypassing the damaged hair cells of the cochlea. The implant helps convert sound into electrical signals. These signals are then sent to an array of electrodes implanted surgically in the cochlea. The implant system preserves the tonotopic map of the cochlea.

Components

- The implant has external components consisting of a microphone which receives sound and transduces it into an electrical waveform, a speech processor which divides the signals into components for each of the electrodes and a transmitting coil which sends the signals across the scalp to the internal components.
- The internal components include a receiver-stimulator which receives the signals from the transmitting coil and sends it to the electrode array which is implanted in the scala tympani of the cochlea.

Selection Criteria

- Bilateral profound cochlear hearing loss unresponsive to amplification by the most powerful hearing aids is the indication for an implant.
- All children below the age of 10 years who have congenital or acquired profound hearing loss and who will not benefit from conventional hearing aids and all adults who have lost hearing after acquisition of language are candidates.
- The only true pre-requisite is an intact auditory nerve.
- The minimum age for implantation in children has come down and children as young as 6 months of age have been implanted. As the cochlea is at full size at birth, there is no anatomic difficulty with electrode insertion in very young children.

Contraindications

- Cochlear aplasia, absence of auditory nerves, retro-cochlear cause of deafness, central deafness, presence of external or middle ear infections and co-existent severe medical illness are contraindications.

Pre-operative Evaluation

- Pre-operative evaluation includes a complete ear, nose and throat (ENT), and head and neck examination, including assessment for additional handicaps, haematological tests, TORCH serology, if required, and skiagram of chest and ECG for assessing fitness for surgery.
- Audiological and electrophysiological investigations include:
  - Pure tone and impedance audiometry otoacoustic emissions (OAE)
  - Brainstem evoked response audiometry (BERA)
  - Auditory steady state response (ASSR)
  - Aided audiometry
  - Hearing aid trial.
- Magnetic resonance imaging is the gold standard for the assessment of cochlear anatomy and the vestibulocochlear bundle. It reveals anomalies like Mondini's and Michel's aplasia, labyrinthitis ossificans and absent eighth nerve.

Surgery of Cochlear Implantation

- The goal of cochlear implant surgery is to insert the entire electrode array into the scala tympani with as little damage as possible to the structure of the inner ear. Surgery is essentially the same for children and adults because the anatomic structures are of adult configuration at birth. However, in very young children, there is a slightly increased risk of facial palsy.

The steps of surgery are:

- ***Incision:*** An extended post-auricular incision to expose the mastoid cortex. The incision made more than 1 cm from the body of the implant.
- ***Simple mastoidectomy:*** The mastoid is drilled out to expose the mastoid antrum.
- ***Posterior tympanotomy:*** The facial recess is opened and the promontory and round window niche are exposed without exposing the facial nerve.
- ***Well for receiver-stimulator:*** This is fashioned in the skull behind the mastoid cavity using a template as a guide and a groove is made to connect it to the mastoid cavity. Tie-down holes are made on either side of the well for securing the implant.
- ***Cochleostomy:*** The basal turn of the cochlea is opened anterior to the round window to make the axis of introduction of the electrode array straighter.
- ***Insertion of electrode array:*** The electrode array is inserted atraumatically into the scala tympani using a claw. Once the electrodes are inserted, diathermy should not be used.
  - Fixation of the device and electrode array and wound closure is done.

Electrophysiologic Testing or Neural Response Telemetry

- Neural response telemetry (NRT) is performed after implanting the electrode array.
- This assures the team that the device is functioning and that the patient is receiving an auditory stimulus and responding appropriately.

Post-operative Care

- The patient is called for review three weeks post-operatively for switch-on of the device.
- Frequent mapping sessions are required and prolonged and intensive rehabilitation after implantation is essential. Rehabilitation aims at improving receptive language skills and expressive skills.

Complications of Cochlear Implantation

- Major complications include facial palsy, and implant exposure due to flap loss and wound infection.
- Other complications include facial nerve stimulation, device failure, deterioration of hearing, tinnitus, temporary balance problems, numbness of the scalp, loss of taste, electrode/device extrusion, CSF leak and meningitis.

AUDITORY BRAINSTEM IMPLANTATION

- Auditory brainstem implant (ABI) is an effective means of hearing rehabilitation in patients with neurofibromatosis type 2.
- In such patients with damaged cochlear nerves on both sides, the brainstem implant bypasses the cochlear nerves and directly stimulates the cochlear nucleus. The cochlear nucleus has tonotopicity.

Indications for Auditory Brainstem Implant

- Multichannel ABIs are indicated for patients with neurofibromatosis type 2 (NF2) and schwannomas involving the internal auditory canal or cerebellopontine angle.

- ABI is being considered for non-tumour patients and even in children with congenital hearing loss before the loss of neuronal plasticity.
- In the near future, it may be used in bilateral temporal bone fractures and demyelinating diseases affecting the eighth cranial nerves, but sparing at least one cochlear nucleus.
- Auditory brainstem implantation has also been used in cases of bilateral totally ossified cochlear in which a cochlear implant cannot be used.
- The current criteria for ABI include evidence of bilateral seventh and eighth cranial nerve tumours involving the IAC or cerebellopontine angle, language competency, age > 12 years or older, psychologic suitability, willingness to comply with research follow-up protocol and realistic expectations.

Pre-operative Evaluation

- A multi-disciplinary approach is essential involving neurotologists, neurosurgeons, audiologists and anaesthetists.
- Prior to planning the surgery, assessment of the tumour and the hearing is vital.
- Comprehensive audiological tests, including pure tone audiometry, brainstem evoked response audiometry, otoacoustic emissions and auditory steady state response (ASSR), are required.
- Pre-operative MRI is very important because it may signal potential problems leading to non-stimulation such as a large lateral recess or tumor damage to the cochlear nucleus region and also helps to rule out other intracranial and spinal tumours.

Auditory Brainstem Implant Surgery

- For successful ABI surgery, a few important issues, such as patient selection, choice of device, choice of approach, technique of tumor removal, knowledge of micro-anatomical variations, intra-operative identification of the cochlear nucleus and prevention of complications, have to be considered.
- Nerve monitoring is required and neuromuscular blocking agents should be avoided during monitoring. EMG monitoring of V, VII and IX nerves is done.
- There are several approaches described for tumour removal and placement of ABI.
  - **Trans-labyrinthine approach** provides optimal access for both removal of the tumour and placement of the electrode array. Disadvantages with trans-labyrinthine approach are limited exposure of cranial nerves and vessels in the posterior fossa.
  - **The lateral suboccipital approach** is preferred by most neurosurgeons as it is fast, safe and offers very good exposure of the lateral posterior fossa.
  - **The middle cranial fossa** approach is only possible with angled endoscopes; however, it is technically the most difficult and places the facial nerve at greatest risk.
- After craniectomy is performed, a seat for receiver-stimulator is created in the area postero-superior to the craniectomy. Tie-down holes are then placed on either side of the receiver-stimulator for securing the implant.
- The cochlear nerve is followed medially as it enters the lateral recess of the fourth ventricle.

- The ABI is inserted in the lateral recess of the fourth ventricle, which is adjacent to both cochlear nuclei.
- Between the bulging of the cochlear nucleus and the ponto-bulbar body, a small straight vein is a constant finding and an important landmark. The typical straight vein at the cochlear nucleus heading to the entrance of the foramen of Luschka is found in 76%.
- The taenia of the choroid plexus at the entrance of the fourth ventricle is dissected and the device is advanced into the lateral recess of the fourth ventricle over the surface of the cochlear nuclei.
- Activation of the device is done 3–6 weeks later.
- **Contraindications to ABI** include previous stereotactic radiotherapy which has the risk of radiation necrosis of the cochlear nucleus region and anatomic distortion and fibrosis. ABI may not be possible in very large tumours which cause distortion of the brainstem.

Auditory Midbrain Implants

- The auditory midbrain implant (AMI) is a new hearing prosthesis designed for stimulation of the auditory midbrain, particularly the inferior colliculus central nucleus (ICC).
- AMIs are placed in the ICC. They may prove to be a safe and potential alternative for hearing restoration in neuro fibromatosis Type 2 (NF2) patients and may help in enhancement in lip-reading capabilities and environmental awareness and some improvement in speech perception performance.

5
CHAPTER

Neuroendocrinology

Ravi Ramamurthi • Ravindranath Kapu

INTRODUCTON

- The anterior pituitary secretes the following hormones:
  - Prolactin (PRL)
  - Growth hormone (GH)
  - Adrenocorticotropic hormone (ACTH)
  - Thyroid-stimulating hormone (TSH)
  - Gonadotrophic hormones which are follicle stimulating hormone (FSH) and luteinising hormone (LH).
- The posterior pituitary secretes: Antidiuretic hormone (ADH or vasopressin) and oxytocin.
- Pituitary hormones are measured by radioimmunoassay (RIA), immunoradiometric assay (IRMA) or enzyme-linked immunosorbent assay (ELISA).
- The reserve capacity of the pituitary gland, in its response to stress, must be tested with provocative tests (dynamic testing).
- Normal basal level listed in Table 1.

Table 1: Normal basal levels

Free Tri-iodothyronine (T3)		2.2–5.0 pg/mL
Free Thyroxine (T4)		0.7–2.2 ng/dL
Thyroid stimulating hormone		ND–9 u IU/mL
17 OH Progesterone		50–180 ng/dL
Progesterone	Post-ovulatory	Above 5 ng/mL
Testosterone	Male	360–990 ng/dL
	Female	15–110 ng/dL
Prolactin	Male	ND–15 ng/mL
	Female	ND–20 ng/mL
Growth hormone	Fasting adults	ND–5 ng/mL
	Fasting children	ND–10 ng/mL
Cortisol	Morning sample	5–25 µg/dL
	Evening sample	2.5–12.5 µg/dL
17 Ketosteroids	Male	9–24 mg/24 hours
	Female	5–17 mg/24 hours
DHEA	Male	80–560 µg/dL
	Female	35–430 µg/dL